

Behcet's Disease: Radiologic Diagnosis

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INTRODUCTION

Behcet's disease is a chronic, relapsing, multisystemic, and inflammatory disease. It causes mucocutaneous, ocular, vascular, articular, gastrointestinal, urogenital, pulmonary, and neurological abnormalities [1-3].

Etiopathogenesis is still a subject of study. Infectious agents, immune mechanisms, and genetic factors are held responsible. However, Behcet's disease is described as a type of systemic vasculitis [1].

The diagnosis is primarily based on clinical criteria because there are no specific diagnostic laboratory tests available and because of the non-specificity of the histopathologic findings. The multiorgan involvement of Behcet's disease can be characterized. Here we will describe the radiological findings consisting of pulmonary, cardiovascular, neurologic, and abdominal involvement in Behcet's disease.

HISTOPATHOLOGY

The underlying histopathologic process in Behcet's disease is inflammation of the vasa vasorum of the tunica media, which causes destruction of the elastic fibers of the media and dilatation of the vessel lumen. Histologic evidence of aortitis can be divided into two stages: active and chronic. Active aortitis include infiltration of inflammatory cells in the media and adventitia and proliferation of the vasa vasorum. Loss of elastic fibers and muscle fibers in the media layer leads to destruction of wall integrity. In the chronic stage of aortitis, fibrous thickening of the intima and adventitia occurs [4]. Weakening of the aortic wall can lead to saccular aneurysm and, less commonly, fusiform aneurysms that may rupture. Perivascular inflammatory cell infiltration leads to occlusion of the vasa vasorum, which in turn leads to transmural necrosis of the walls of large muscular arteries. Saccular aneurysms are probably produced by severe destruction of elastic fibers in the media and consequent perforation of the vessel wall [4,5]. Thickening of the vessel wall is caused by inflammation and infiltration by lymphocytes, plasma cells, and neutrophils [3].

Pulmonary Involvement

Mediastinal lymphadenopathy may be seen and is probably a reaction to an inflammatory process in the thorax.

Lung parenchyma

Actual prevalence of pulmonary alterations (**PAs**) is unknown in Behcet's disease because there have been no prospective studies evaluating pulmonary symptoms in randomly selected patient groups [1].

The PAs that are seen in Behcet's disease are nonspecific changes and could arise from various causes such as inflammation, pneumoconiosis, pulmonary infarct, and so on [1].

The reported prevalence of pulmonary involvement of Behcet's disease ranges from 12% to 16% [1,6]. The pulmonary alterations associated with Behcet's disease include pulmonary infarcts, pulmonary hemorrhage, atelectasis, fibrosis, emphysema, wedge-shaped or linear shadows, undefined nodular or reticular opacities due to cryptogenic organizing pneumonia, eosinophilic pneumonia, recurrent pneumonia, bronchitis, patients with or without pulmonary artery aneurysms and chronic pulmonary thromboembolism [1,6-8].

Wedge-shaped or ill-defined increased opacities with rapid resolution are considered pulmonary infarction, recurrent pneumonia, hemorrhages due to pulmonary artery aneurysm (Figure 1). In chronic pulmonary thromboembolism, lung parenchyma shows mosaic attenuation. Airspace consolidation can also result from infectious pneumonia, which might be recurrent. Pneumonias in Behcet's disease can be the result of inflammation in the vessels of pulmonary parenchymal, which may occur secondary to immunosuppressive therapy [8]. Damaged lung tissue can be replaced by fibrosis or emphysema. Possible causes for hemoptysis include rupture of an aneurysm with erosion into a bronchus [8,9].

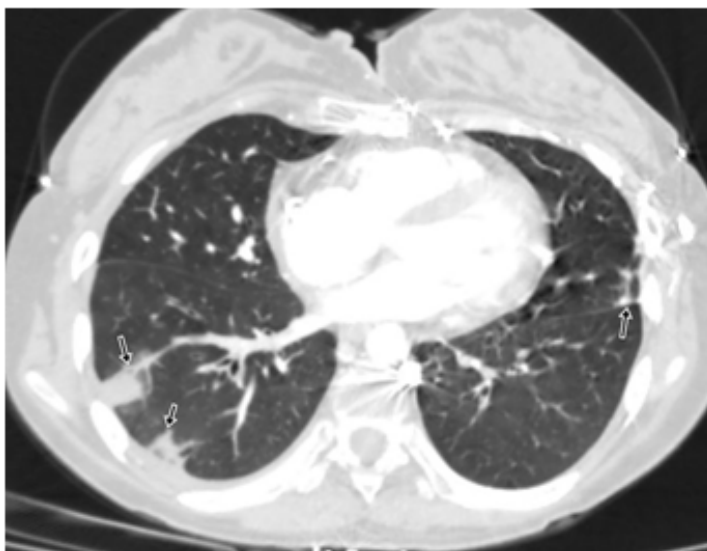


Figure 1: Vasculitis with hemorrhage and infarction in a 33-year-old patient. Chest CT scans (lung window) obtained at middle level of the right lower lobe demonstrate focal areas of high attenuation in the periphery of the lobe (arrows), findings that represent vasculitis with hemorrhage and infarction.

Pleura

Vasculitis of the pleura may result in the formation of pleural nodules, which are often difficult to differentiate from subpleural parenchymal lesions (Figure 2). Pleural effusion may be imputed to pulmonary infarction, vasculitis of the pleura, or superior vena cava SVC thrombosis [10]. Also, pericardial effusion may be attributed to vasculitis of the pericardium or SVC thrombosis.



Figure 2: Vasculitis of the pleura in a 32-year-old patient. Chest CT scan (mediastinal window) demonstrates tiny pleural nodules (arrow) and pleural fluid (thick arrow), in the right lower lobe findings being consistent with vasculitis of the pleura.

Cardiovascular Involvement

Cardiac Involvement

Cardiac involvement was present in 1% 5% of Behcet's disease patients seen in a previous clinical series [11] and in 16.5% of cases in the Japanese autopsy registry [12]. Although the frequency of cardiac alterations is minimal, they can lead to serious clinical conditions. Cardiac alterations associated with Behcet's disease include intracardiac thrombosis, endomyocardial fibrosis, periaortic pseudoaneurysm, rupture of the aortic sinus, endocarditis, myocarditis, pericarditis, pericardial effusion, coronary artery disease, myocardial infarction, aneurysm of the aortic sinus, valve dysfunction, and conduction system disturbances [13,14].

Intracardiac thrombosis and endomyocardial fibrosis: Intracardiac thrombosis has been observed in patients with Behcet's disease, with the right side of the heart being the most frequent site of involvement [15] (Figure 3).



Endomyocardial fibrosis is a rare manifestation of cardiac involvement in Behcet's disease. Vasculitis that involves the endocardium, myocardium, or both may be complicated by intraventricular thrombus [11]. Imaging findings consist of an intraventricular filling defect with or without calcification, dense fibrous tissue with inflammatory infiltrates and numerous vessels in the endocardium, bright echo at echocardiography or low computerized tomography (CT) attenuation along the myocardium, displacement of leaflets of the tricuspid or mitral valve, and right atrial enlargement with or without narrowing of the ventricles.

Aneurysm and rupture of the aortic sinus: Aneurysms of the aortic sinus can be either congenital or acquired, although the majority of cases are congenital. Aneurysms of the coronary arteries have also been described. The majority (94%) of aneurysms of the aortic sinus originate from the right coronary sinus, while only 6% originate from the noncoronary sinus. Almost all such

aneurysms project into the right atrium or right ventricle [15,16]. Active inflammation of the aorta may account for the rupturing of an aneurysm. It is difficult to diagnose an unruptured aneurysm of the aortic sinus, as almost all patients are asymptomatic. Transesophageal echocardiography is more useful than transthoracic echocardiography for making this diagnosis. Cardiac magnetic resonance imaging (**MRI**) can easily depict an aneurysm, and cine MRI sequences are especially useful for demonstrating aneurysm rupture into the cardiac chambers.

Vascular involvement

If Behcet's disease is associated with lesions in the large vessels, it is referred to as "vasculo-Behcet disease."

Although vascular involvement is seen in only 25% of patients, it is the most common cause of mortality in Behcet's disease [17]. Systemic arterial alterations of Behcet's disease are infrequent compared with venous involvement, accounting for only 12% of vascular complications [9,18]. Vascular alterations of Behcet's disease in the thorax may be seen in the aorta, pulmonary artery, pulmonary vein, brachiocephalic artery, SVC, and brachiocephalic vein, in the superior vena cava. Vascular involvement of Behcet's disease can be divided into three subsets including venous occlusion, arterial occlusion, and arterial aneurysm. Identification of a vascular lesion is important because it seriously affects a patient's prognosis. The leading cause of death in patients with Behcet's disease is the rupture of a large aortic or arterial aneurysm [17].

Venous occlusion: Deep and superficial veins thrombophlebitis is the most common form of venous occlusion in Behcet's disease [17]. Superior vena cava (**SVC**) thrombosis is not uncommon (Figure 4), [19] and is often accompanied by thrombosis of other mediastinal veins. SVC syndrome caused by thrombotic occlusion of the SVC and the accompanying fibrosing mediastinitis results in the development of mediastinal collateral veins in the neck and chest wall, the esophageal veins and into the coronary vein, hepatic vein, and inferior vena cava (**IVC**), giving rise to a downhill varix [20] (Figure 5). Venous occlusion is the most common pathology, followed by arterial aneurysm and arterial occlusion. Thrombosis actually begins in the SVC or adjacent large veins without continuation between them. CT can reveal mediastinal widening, intraluminal filling defects in the SVC, wall thickening and obliteration of the SVC with soft-tissue edema, and infiltration into the mediastinal fat. Thrombosis of the SVC accompanying fibrosing mediastinitis is one of the unique features of Behcet's disease [20]. Deep vein thrombosis in the lower extremities is also a frequent finding, along with thrombosis in the SVC, IVC, and veins in the upper extremities (axillary, brachial, etc.), (Figure 6).



Figure 4: SVC thrombus in a 39-year-old patient. Contrast-enhanced coronal chest CT scan (mediastinal window) demonstrates a thrombus SVC (arrows).

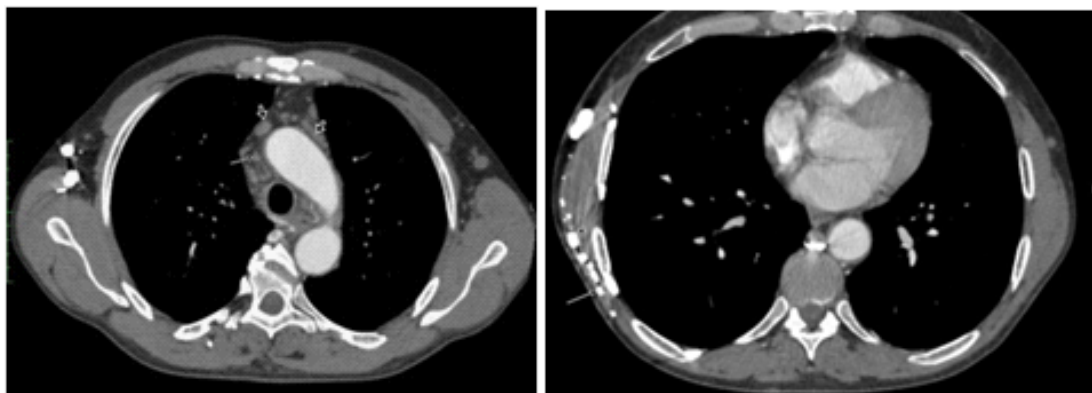


Figure 5: a, b. SVC thrombosis and collaterals in a 55-year-old patient.(a) Contrast-enhanced chest CT scan (mediastinal window) demonstrates a narrow and fibrotic SVC (arrow) and extensive collateral vessels in the mediastinum (thick arrows). (b) Contrast-enhanced chest CT scan (mediastinal window) demonstrates extensive collateral vessels in chest Wall (arrows).

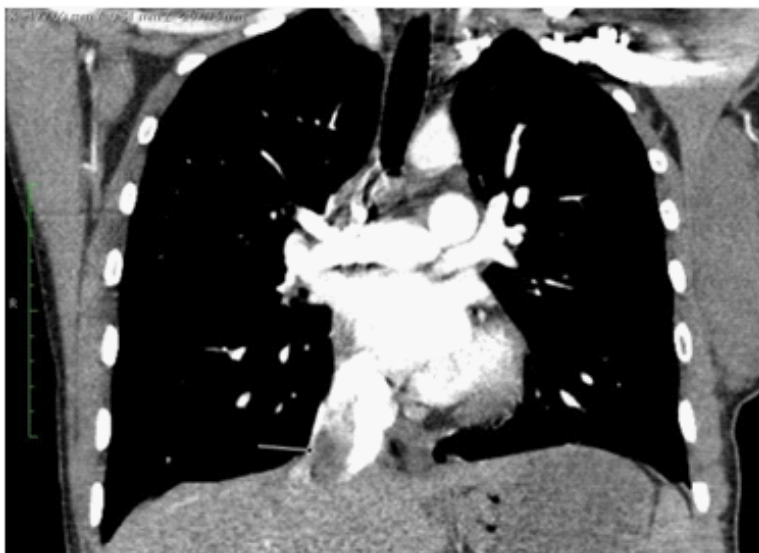


Figure 6: IVC thrombosis in a 19-year-old patient. Contrast-enhanced coronal chest CT scan (mediastinal window) demonstrates a thrombus IVC (arrow).

Arterial occlusion: Arterial occlusions rarely occur and represent 1.5% of all forms of vascular involvement in Behcet's disease [5]. However, one large study of vascular Behcet's disease showed a higher frequency of arterial occlusion than of arterial aneurysm [21]. In that series, aneurysms were common in large and medium-size arteries, while occlusions or stenoses were common in the distal runoff arteries of the lower extremities [21]. The authors concluded that the lumen of smaller arteries is often obliterated during the active stage of vasculitis because of the intense infiltration of inflammatory cells in the media and adventitia, swelling of the intima, and thrombosis in the lumen.

Arterial aneurysm: The abdominal aorta is the most frequent site of arterial involvement of aneurysms, and the pulmonary artery is the second most common site, followed by the coronary arteries, aortic arch, femoral, subclavian, and popliteal arteries [5,21],(Figure 7).

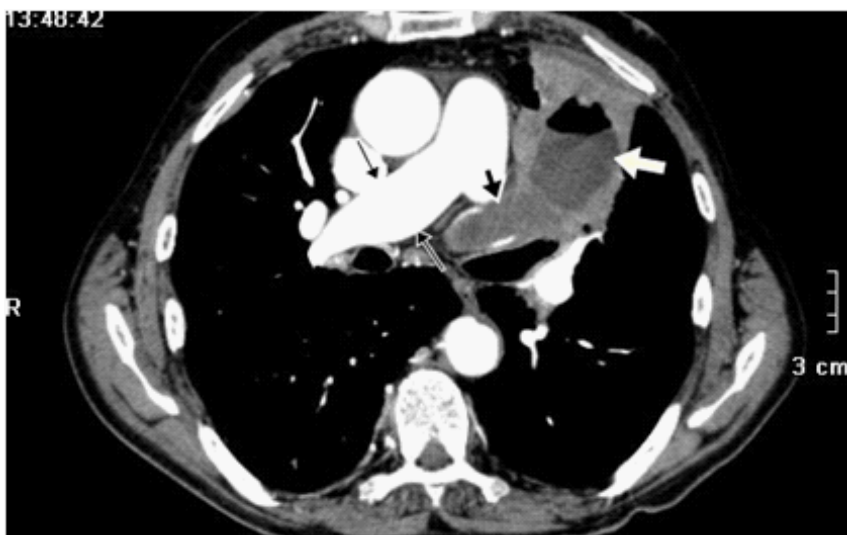


Figure 7: Pulmonary artery aneurysm, pulmonary vein thrombus and pulmonary abscess in a 65-year-old patient. Axial contrast-enhanced chest CT scan (mediastinal window) demonstrates an aneurysm of the right main pulmonary artery (arrows), thrombus in left pulmonary vein (thick arrow), and abce in inferior lingular segment (white arrow) .

Behcet's disease is the most common cause of pulmonary artery aneurysm [22]. Aneurysms are usually pseudoaneurysms and are often accompanied by varying degrees of mural thrombus. They are most frequently located in the right lower lobe artery, but may also appear in the right and left main pulmonary arteries [23] (Figure 8). Bilateral involvement is not rare, and aneurysm diameters vary. The most frequent symptom is hemoptysis with ruptured aneurysm erosion into a bronchus [23,24]. Aneurysm formation in the pulmonary arteries indicates a poor prognosis: 30% of patients with this condition will die within 2 years [3,25,26].

Neurologic Involvement

Behcet's disease is classified into two main groups defined as parenchymal and non-parenchymal CNS involvement [27]. Primary progressive, secondary progressive and relapsing-remitting forms are also defined. Cranial and spinal MRI, MR venography or cerebral angiography, should be performed for all suspected neuro-Behcet's disease (NBD) patients.

Parenchymal (intra-axial) Involvement

Parenchymal involvement is usually seen in the cerebral hemispheres, brainstem, pyramidal tracts, and spinal cord. In most patients, depending on the small venous inflammatory disease of the central nervous system, focal or multifocal parenchymal lesions are seen (Figures 8-12), [28]. Lesions of the brain stem and basal ganglia usually extend to the diencephalic structures (Figure 9). Lesions in the brain stem can be isolated and often are located in the pons (Figure 10).

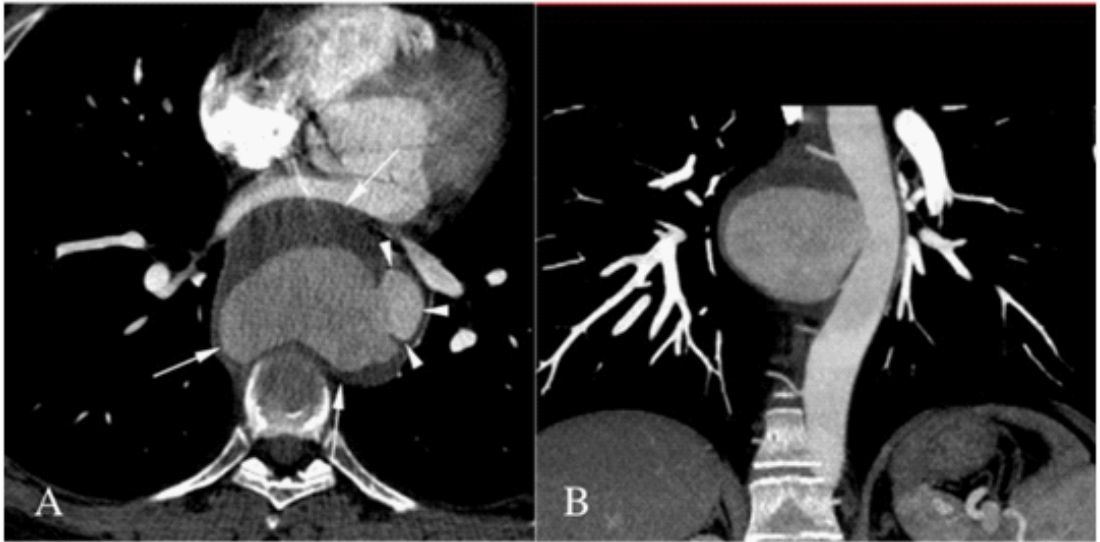


Figure 8: Thoracic aorta aneurysm in a 33 year old patient. Contrast-enhanced (A) axial, (B) coronal chest CT scan (mediastinal window) demonstrates a huge aneurysm (arrow) originated from right side of thoracic aorta (arrowheads) and displacement of the heart anteriorly. Thickened aneurysmal wall with partial thrombosis is seen.

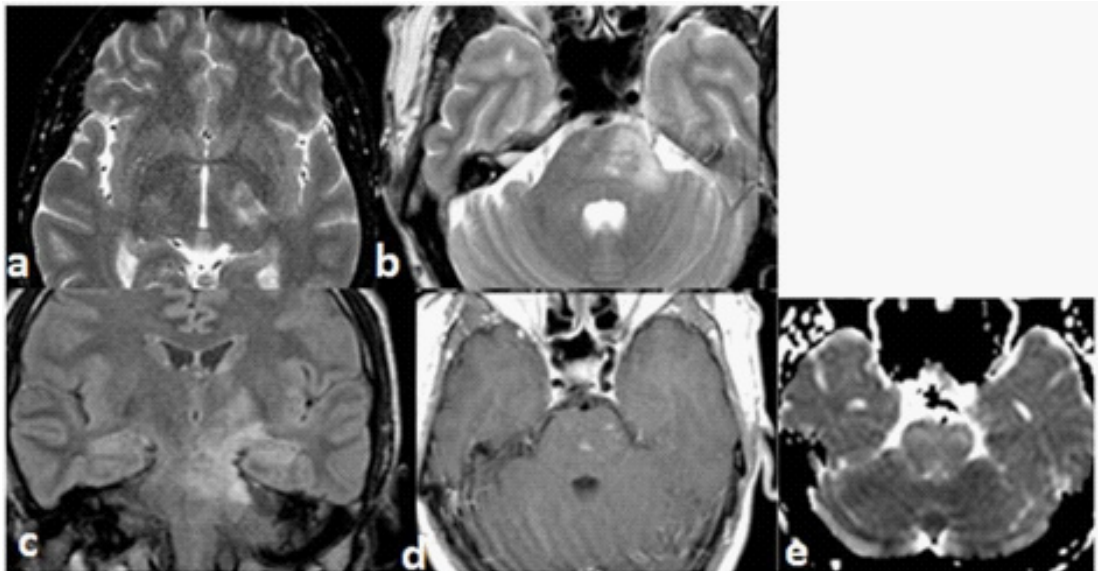


Figure 9: Basal ganglia and brain stem involvement during an acute stage of NBD. T2W axial (a, b) and flair coronal (c) images show that large hyperintense lesion area extending between the left posterior limb of internal capsule, thalamus, midbrain and pons. Punctate enhancement is seen only in a small part of this large lesion on the contrast-enhanced axial T1W image (d). Increased diffusion is seen on ADC map image (e).

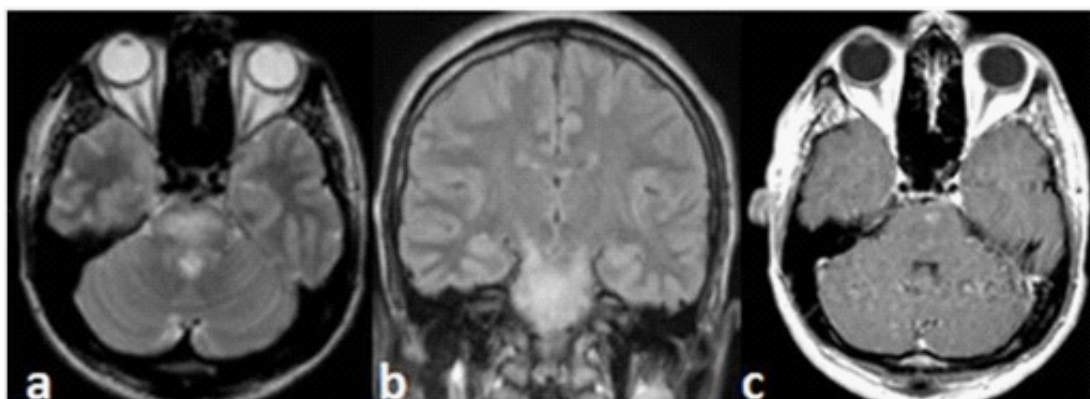


Figure 10: Cranial MRI during an acute attack of neuro-Bechet's disease. T2W axial (a), flair coronal (b) images show hyperintense lesion, which is completely covering the pons and extending to the midbrain. Only partial contrast enhancement is seen axial contrast-enhanced T1W image (c).



Figure 11: Spinal Behcet's disease. Patchy hyperintense medullary lesions (short arrow) are seen at the C2, C4, and C5 levels on the T2W sagittal image (a). However, enhancement of medullary lesion is seen only at the C2 level (long arrow).

Following the treatment period the lesions generally shrink and remain as millimetric T2 hyperintensities. Later the lesions often disappear or become smaller and unenhanced [29]. Significant brain stem atrophy may develop especially after repeated attacks in the late period. Although there is some correlation between detected lesions on MRI and neurological symptoms, the lesions may be larger than expected with common concomitant peripheral edema.

Diffusion-weighted imaging (**DWI**) is a useful tool to differentiate lesions from acute infarction [30]. Slight hyperintensity on DWI with no evidence of diffusion restriction on apparent diffusion coefficient (**ADC**) maps can be seen in case of acute exacerbation of NBD patients (Figure 9).

Approximately 10% of NBD patients have white matter lesions on MRI. Although these may be due to nonspecific causes, multiple sclerosis should be kept in mind [31]. The differential diagnosis of NBD includes systemic lupus erythematosus, sarcoidosis and tumoral processes [27]. Advanced techniques such as MR spectroscopy and perfusion MR (pMRI) may be useful in the differential diagnosis of tumors. Rarely, stereotactic biopsy may be required [32].

Nonparenchymal (extra-axial) Involvement

The primary problem of non-parenchymal CNS involvement is not associated with CNS parenchyma. It is more preferably related with neurological signs of brain venous or arterial involvement. In addition, subjects with only leptomeningeal or dural involvement with normal brain parenchyma should also be included in this group.

Dural sinus thrombosis, bilateral vertebral or internal carotid artery occlusion, and aneurysms are considered in this group [29,33]. Cerebral venous sinus thrombosis is seen in 10-20% patients with extra-axial Behcet's disease. Patients show signs of intracranial hypertension and papilledema. One or two-sided sixth nerve palsies can occur as well as papilledema in these patients while parenchymal CNS involvement is practically never seen [34,35]. Non-parenchymal CNS involvement has better prognosis compared parenchymal NBD. Cranial MRI and MR venography are the first noninvasive imaging techniques that should be elected for suspected cases. The diagnostic value of these test is higher when they are used together and digital subtraction angiography is rarely needed. The most common site for thrombus is the superior sagittal sinus, usually accompanied by lateral sinus thrombosis. In the acute period (1-5 days) the occluded sinus is seen as isointense on T1-weighted (**T1W**) and hypointense on T2-weighted images (**T2W**). It appears hyperintense on T1 and T2-W images in the subacute phase (Figures 12,13). CNS arterial involvement may occur rarely in NBD presenting with stroke-like syndrome or arterial aneurysm [35].

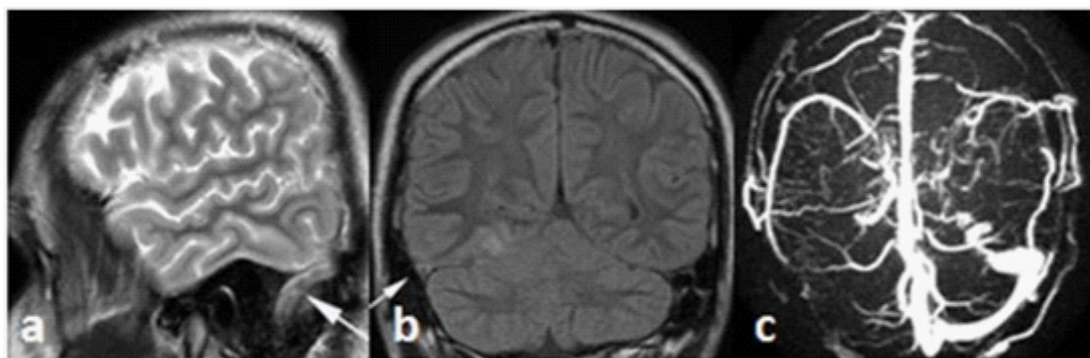


Figure 12: Transverse sinus thrombosis in Behcet's disease. Thrombus hyperintensity in the right transverse sinus (arrow) is seen at sagittal T2W (a) and flair coronal images. Abscense of flow in the right transverse and sigmoid sinuses is seen the phase contrast in MR angiography MIP image (c).

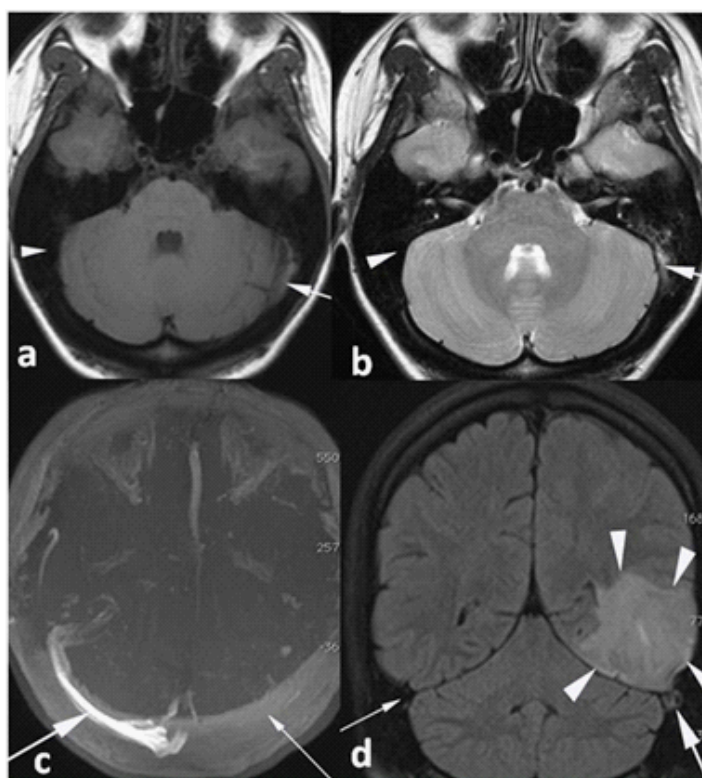


Figure 13: Venous sinus thrombosis and venous infarction in a patient with Behcet's disease. Axial T1W (a) and T2W (b) images shows that hyperintense thrombosis (arrow) in the left sigmoid and transverse sinus. 3D TOF MR angiography image (c) shows that the absense of flow in the the left transverse sinus (thin arrow). Large swollen venous infarction is seen in the brain parenchyma (arrowheads) adjacent to left sigmoid thrombosis (thick arrow).

In the literature, several advanced imaging techniques have been applied to NBD patients. A current study using (99m) Tc-HMPAO brain SPECT scans showed impaired perfusion especially in the temporal lobes in all NBD patients with neurological complaints (24 out of 24, 100%) and normal brain MRI and CT findings [36]. In another study including patients with brain involvement or non NBD, pMRI was found to be a very sensitive method to detect brain involvement in NBD patients with negative MRI [37].

Orbital and Spinal Involvement

Spinal cord involvement in NBD patients may be seen alone or together with brainstem involvement [29,31]. Coexistence with brainstem involvement leads to more severe clinical presentation. Single or multiple level T2-hyperintense focus of the spinal cord can be seen on spinal MRI (Figure 11), [38]. As a manifestation of NBD ocular involvement and, rarely, optic neuritis may be seen on MRI [39]. MR images show a thickened optic nerve, and contrast-enhanced posterior scleral area (Figure 14).

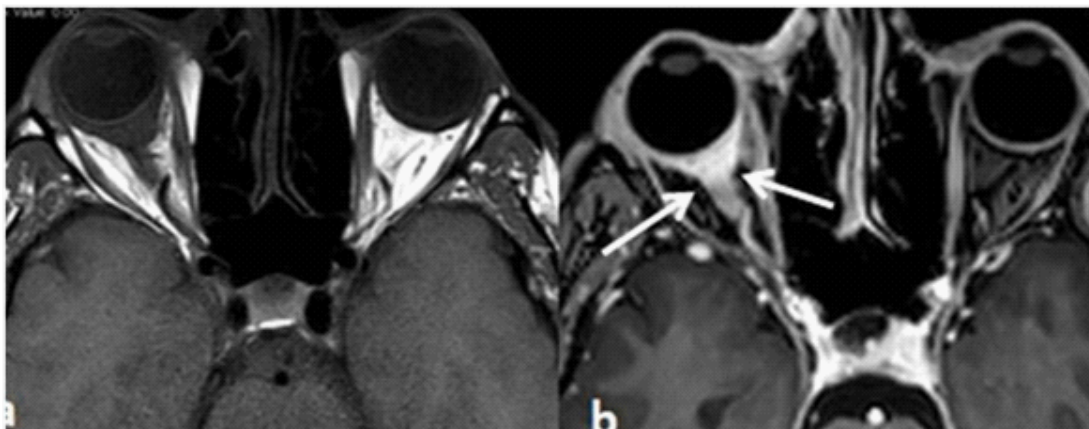


Figure 14: Orbital involvement in a patient with Behcet's disease. Unenhanced (a) and contrast-enhanced (b) axial T1W images shows the thickened and enhanced right optic nerve and posterior scleral area (arrows).

Behcet's Disease In Abdomen

Behcet's disease is a chronic multisystemic recurrent inflammatory disease that involves vessels of all sizes in various organs and is characterized by non-specific histopathologic vasculitis [40,41]. In this chapter, we focus on gastrointestinal and abdominal vascular involvement.

Gastrointestinal involvement: Behcet's disease involves the gastrointestinal system in 5-60% of patients, depending on the geographic region in the world [42]. It has a wide variety of clinical symptoms including loss of appetite, abdominal pain, bloody diarrhea, nausea, vomiting, dysphagia, and abdominal distension [43]. Intestinal lesions including mucosal inflammations, ulcerations, and ischemic damage result from neutrophilic phlebitis and vasculitis [45]. The

ileocecal area is the most frequently involved region; however, any part of the gastrointestinal tract and extra-intestinal organs including spleen, pancreas or liver can be involved [44,45].

Esophagus: Esophageal involvement is rare, with an incidence of 2-11%; when it is involved, it is often associated with involvement of another part of the gastrointestinal tract [46,47]. It usually involves the middle part of the esophagus. Clinical symptoms include retrosternal chest pain, odynophagia, dysphagia, melena, and hematemesis [48]. Lesions usually appear as single or multiple erosions or ulcerations, diffuse esophagitis, mucosal dissection, intramural hematoma, and stenosis [47,49]. Involvement of the esophagus may lead to perforation and formation of esophagobronchial or esophagotracheal fistula [47]. Histopathological examination reveals non-specific lymphocytic or neutrophilic infiltration rather than vasculitis [46]. These findings are not specific to esophageal involvement of Behcet's disease. Some of these findings can be demonstrated radiologically with double-contrast examinations of the esophagus. CT shows esophageal wall thickening due to edema and inflammation and extraluminal signs accompanying complicating situations. Esophageal varices can be seen in Behcet's disease as a result of SVC obstruction and defined as downhill varices [50].

Stomach and Duodenum: The stomach is the least-involved segment of the gastrointestinal tract. The most common symptoms are dyspepsia and epigastric abdominal pain. Aphthous ulcers are the most frequent gastric and duodenal lesions in Behcet's disease [12,51,52]. These ulcers are not specific for Behcet's disease and can be seen in various diseases that affect the stomach and duodenum. Dieulafoy's lesions and gastric non-Hodgkin's lymphoma involvement of the stomach are rare manifestations [53,54]. Imaging of superficial ulcers is difficult by double contrast examination. Deep ulcers are seen as well-demarcated ulcers in the stomach and duodenum by barium studies [44]. A characteristic radiological finding in Behcet's disease is pyloric stenosis in the absence of the duodenal deformity [44,51].

Small intestine and ileocecal region: The ileocecal region is the most commonly involved site. However, only the terminal ileum can be involved; the antimesenteric side of the terminal ileum is most commonly affected [42,52,55]. The ulcers may be aphthous or deep and round with a punched-out appearance. Longitudinal ulcers are rare. The main sign of Behcet's disease is the presence of large, deeply penetrating ulcerations of the submucosa, muscle layer, or entire intestinal wall [52]. Therefore, there is a high prevalence of complications including perforation, hemorrhage, fistula, and peritonitis [41,52].

A large ovoid or irregular ulcer with marked thickening of the surrounding intestinal wall at the sites of involvement is determined by barium examination. In some cases, small, multiple, discrete, "punched-out" ulcers with considerable thickening of the surrounding mucosal folds can be seen [42]. The lesions cannot be determined when they are small in size or with low quality small-bowel follow-through images. Enteroclysis is more sensitive for determining lesions but it is an invasive procedure. Intestinal Behcet's disease can be diagnosed by showing aphthous

ulcers and pseudopolypoid lesions via capsule endoscopy [56]. Therefore, barium examination is replaced by endoscopic investigation for mucosal lesions.

In the case of lesions penetrating through the bowel wall, endoscopic and barium studies have limitations. CT and MRI are proved to be effective for detecting both the involved bowel segment and extraluminal complications. CT and MRI scans shows concentric or asymmetric bowel-wall thickening that markedly contrast agent enhancement (Figure 15). This enhancement is caused by the stasis of blood due to vasculitis and perivasculitis of the veins and venules of the submucosa surrounding the ulcers [42,46,57]. Perienteric infiltration, lymphadenopathy and mesenteric vascular dilatation may also be seen. The presence of severe perienteric infiltration increases complication risks such as microperforation or localized peritonitis [57].

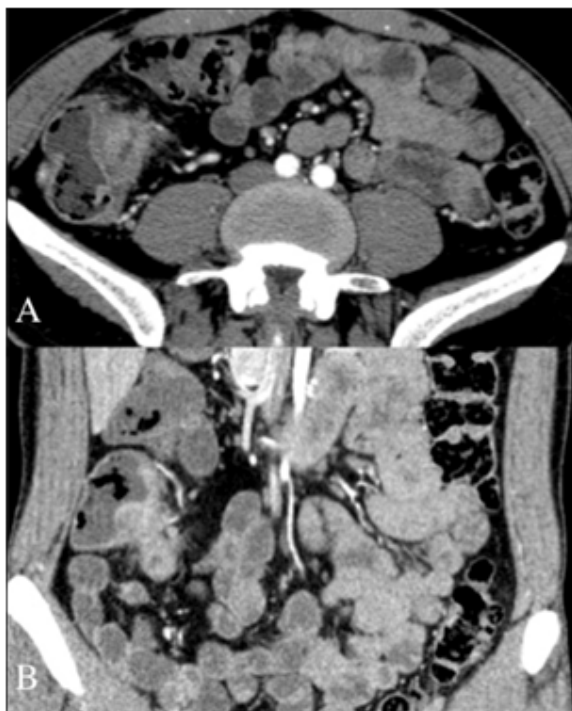


Figure 15: Intestinal involvement in a 35-year-old male patient with Behcet's disease. A deep punched ulcer in the terminal ileum was detected by endoscopic study. A. and B. Contrast-enhanced axial CT images show concentric bowel-wall thickening and wall enhancement in the ileocecal region. Mesenteric lymph nodes and perienteric infiltration are also seen.

Large bowel: The ascending colon is the most commonly involved site in Behcet's disease; however, any part of the large intestine can be involved in Behcet's disease [42]. Unlike ileocecal ulcers-which appear as localized and deeply penetrating-multiple, discrete and punched-out ulcers are commonly seen diffusely in the colon [41].

Differential diagnosis: Differential diagnosis of intestinal Behcet's disease includes Crohn's disease, ulcerative colitis, cecal tuberculosis, and malignant or benign tumors of the cecal area. Both Crohn's and intestinal Behcet's disease manifest as discrete ulcers and discontinuous bowel involvement with relative sparing of the rectum [44]. Longitudinal ulcers, a cobblestone appearance, stricture, fistula, and abscess formation are identified frequently in Crohn's disease [43,45], in contrast to cases of intestinal Behcet's disease [44]. Larger and deeper ulcers in the absence of granuloma formation and the common occurrence of bowel perforation are more common in intestinal Behcet's disease [42,52]. Thickening and enhancing of the bowel wall are seen by CT and MRI for both intestinal Behcet's disease and Crohn's disease. In addition, pseudosacculations resulting from relative sparing of the antimesenteric border within an affected bowel segment, fibrofatty proliferation around involved bowel loops, and the comb sign corresponding to increased mesenteric vascularity are characteristic CT and MRI signs of Crohn's disease (Figure 16). However, all these findings cannot be seen simultaneously in patients with Crohn's disease. Nevertheless, diagnosis based on radiologic imaging is difficult and requires careful evaluation of clinical findings.

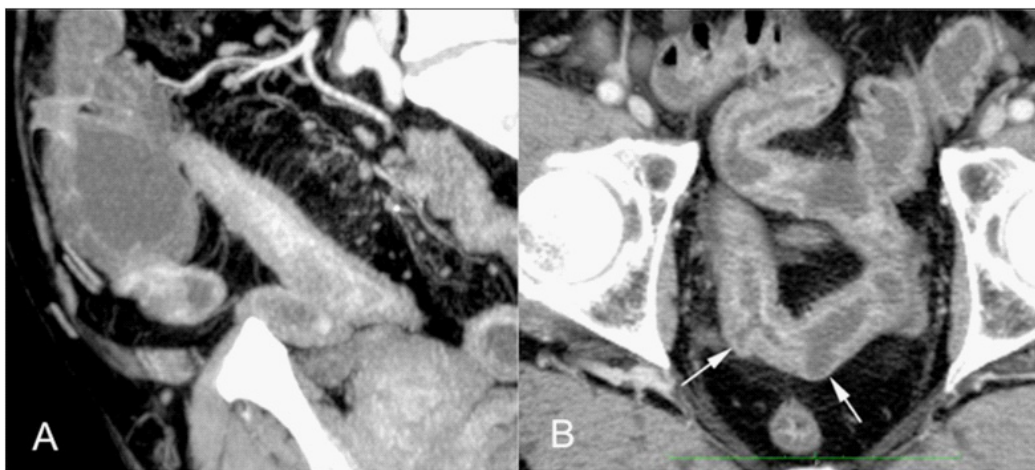


Figure 16: Ileal involvement of Chron's diseases. CT scan shows wall thickening and enhancement in the distal and terminal ileum. A. Prominently dilated adjacent mesenteric vessels (Comb sign), lymph nodes and B. pseudosacculations (arrows), results from relative sparing of the antimesenteric border within an involved bowel loops, are also seen.

Behcet's disease usually involves the ileocecal region or proximal ascending colon with a low rate of rectal involvement-unlike ulcerative colitis, which starts at the rectum and moves to the right colon [43,44,52]. Colonic Behcet's disease appears as one or multiple aphthous or small ulcers with the preservation of the haustra apparent on endoscopic examination, a finding that is rare in cases of ulcerative colitis [44].

Intestinal tuberculosis is usually associated with abdominal pain, fever, and weight loss. The diagnosis is challenging in the absence of pulmonary tuberculosis. The most frequent involvement

in intestinal tuberculosis is seen in the ileocecal region due to the presence of abundant lymphoid tissue. Skip lesions are rare in contrast to intestinal Behcet's disease and Crohn's disease [44]. The radiologic findings of intestinal tuberculosis are bowel-wall thickening, intramural masses, strictures, and enlarged lymph nodes (Figure 17). Peripherally enhancing or calcified lymph nodes may also be seen.

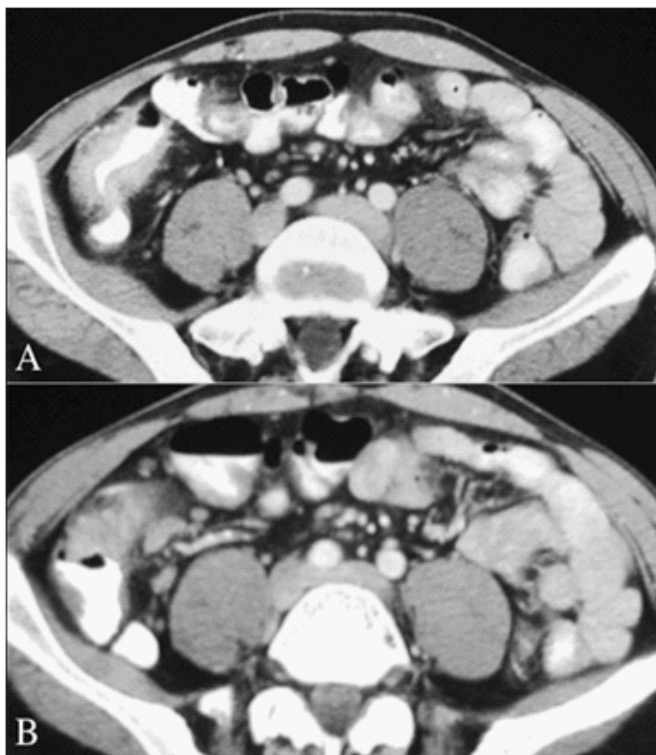


Figure 17: Intestinal tuberculosis involving the terminal ileum in a 25-year-old male patient.

A. and B. Contrast-enhanced axial CT scans demonstrate wall enhancement and concentric wall thickening in the terminal ileum with mesenteric lymph nodes around the involved bowel segment.

Vascular Involvement in Abdomen

The frequency of vascular involvement in Behcet's disease ranges from 5-30% of patients [5,21]. Vascular involvement can be venous or arterial.

Venous Involvement

Endothelial dysfunction secondary to vasculitis is thought to be the cause of venous thrombosis [58][59]. The most frequent involvement is seen as superficial thrombophlebitis followed by deep vein thrombosis in the lower extremities. Thrombosis is seen most frequently in SVC, followed by the inferior vena cava (IVC) and veins in the upper extremities [41].

Hepatic vein and IVC thrombosis: Thrombosis within the hepatic vein and/or the hepatic or suprahepatic segment of the IVC leading to Budd-Chiari syndrome is an unusual disorder. The prevalence of Budd-Chiari syndrome in Behcet's disease has been reported as 3.2% [59]. Behcet's disease entails a high risk of complications and death due to portal hypertension and liver failure. Signs and symptoms of Behcet's disease include right-upper-quadrant abdominal pain, hepatosplenomegaly, and ascites [58].

Hepatic vein thrombosis can be demonstrated by imaging methods such as Doppler ultrasonography (**US**), CT, or MRI. On Doppler US, thrombus within hepatic veins and abnormal collateral vessels that are heading toward the surface of the liver may be seen. It can be difficult to visualize stenotic-occlusive veins in chronic stages; MRI and CT can better visualize thrombosis within hepatic veins and the hepatic parenchyma. They also demonstrate areas of reduced perfusion or necrosis. CT and MRI findings in Budd-Chiari syndrome vary according to whether the condition is acute or chronic. In acute Budd-Chiari syndrome, there are the hepatomegaly associated with congestion, lack of opacification of the hepatic veins, ascites, and absence of collaterals (Figure 18). Splenic enlargement is unusual in the acute phase [60]. The liver appears to be inhomogeneous with a mottled appearance on contrast imaging. The peripheral zones of the liver may appear hypoattenuating and show delayed enhancement due to reversed portal venous blood flow associated with increased postsinusoidal pressure resulting from hepatic venous obstruction [60]. In chronic stages, peripheral atrophy of the liver is seen in Budd-Chiari syndrome but the caudate lobe tends to be spared or enlarged because veins drain directly toward the IVC [60]. Collateral circulatory pathways, including intrahepatic, systemic, and portosystemic vessels are seen, as is splenomegaly associated with portal hypertension (Figure 19). In the chronic stage of IVC thrombosis that does not extend toward hepatic or suprahepatic segments, extensive systemic collaterals are also developed (Figure 20).

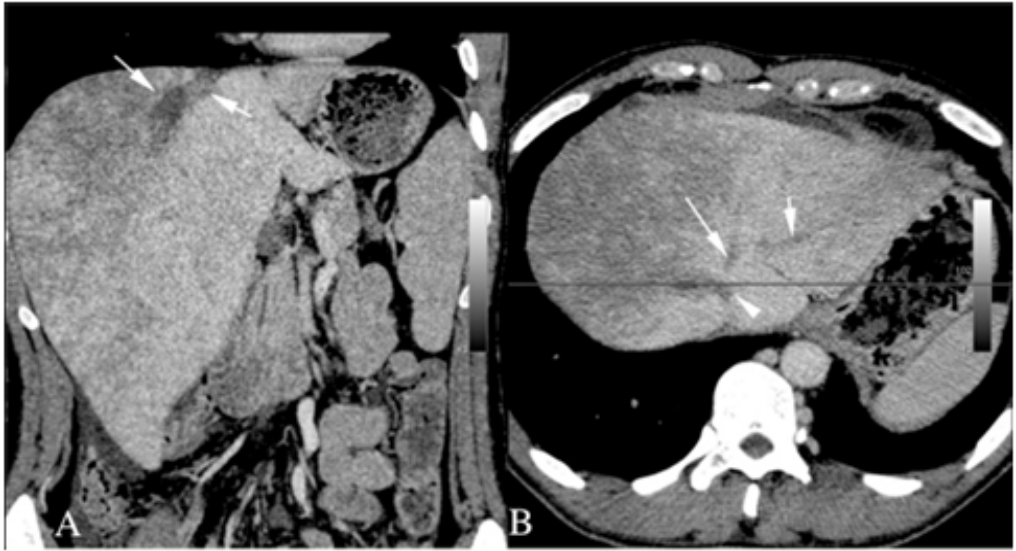


Figure 18: A 32-year-old male patient with Budd-Chiari syndrome in BD. A. Coronal CT scan demonstrates subacute thrombosis seen as low-attenuated complete filling defect in the right hepatic vein. B. Contrast-enhanced axial CT scan shows chronic thrombosis seen as filling defect in the narrowed left and middle hepatic veins. The enhancement of liver parenchyma is inhomogeneous; some areas show lower attenuation than others due to hepatic congestion and necrosis.

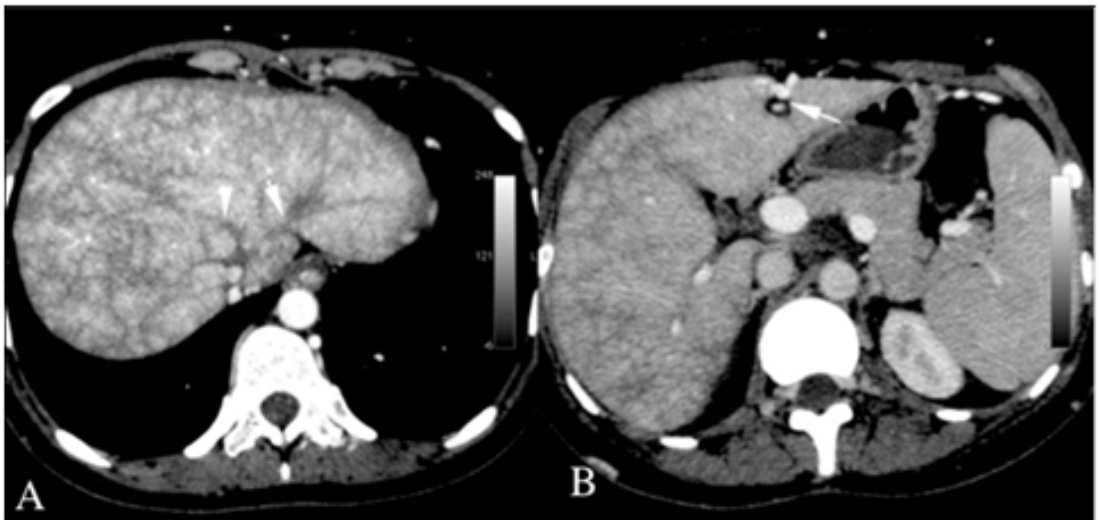


Figure 19: A 28-year-old female patient with chronic stage Budd-Chiari syndrome in BD. A. and B. Contrast-enhanced axial CT images demonstrate chronic occlusion of left and middle hepatic veins (arrowheads) and inhomogeneous, mottled mosaic pattern of parenchymal contrast enhancement. Paraumbilical collateral veins (arrow) and splenomegaly related to portal hypertension are also seen.

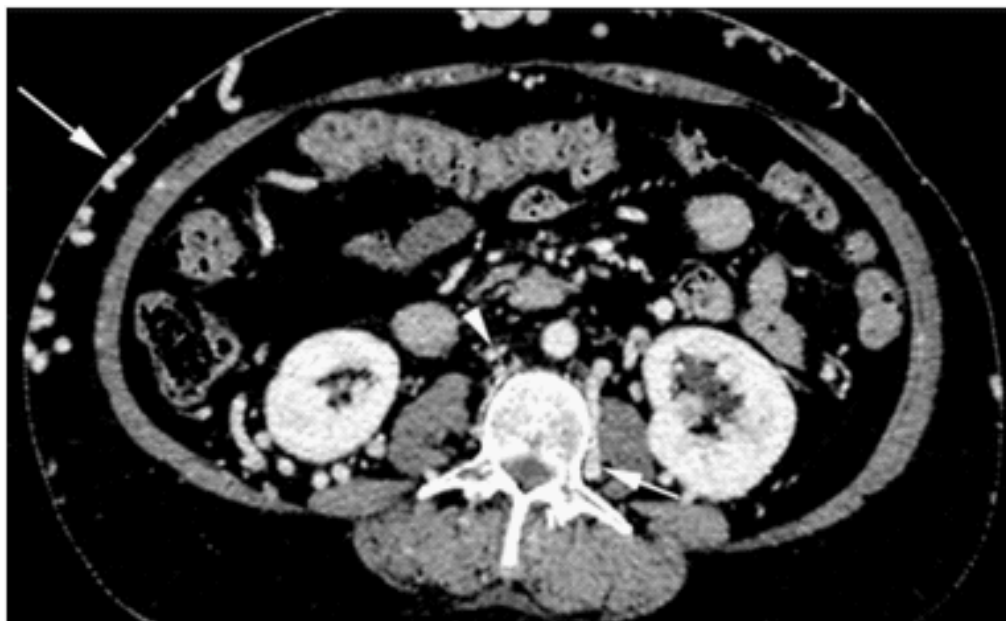


Figure 20: A 52-year-old female patient with chronic IVC thrombosis in Behcet's disease. Axial CT image shows a narrowed IVC (arrowhead) leading to pericaval, paraspinal (arrow), retroperitoneal, and abdominal wall collateral formation.

Portal vein thrombosis: Portal vein involvement is seen in 9.1% of patients with Behcet's disease [61]. Portal vein involvement can appear with either acute or chronic thrombosis. In the acute stage, thrombosis is seen as a filling defect in the lumen without collateral formation, whereas in chronic stage it is seen as cavernous transformation with multiple tortuous vascular channels along the route of the thrombosed portal vein in the liver hilus (Figure 21). Patients with portal vein involvement may exhibit venous infarction in liver parenchyma, especially in the presence of hepatic venous thrombosis [41,58].

Arterial involvement: Arterial involvement of the intra-abdominal organs is rare and presents as aneurysms or occlusion [41,62]. The most frequent site of aneurysms is the abdominal aorta [5,21]. Patients may present with fever, abdominal pain, or a pulsatile mass in arterial involvement. Complications can include intestinal or extra-intestinal organ infarction and gastrointestinal hemorrhage [58]. Involvement of visceral arteries is very rare. There are sporadic case reports in the literature regarding aneurysm formation related to involvement of the celiac trunk, hepatic, splenic, inferior mesenteric, and ileocolic arteries [58]. In addition, arteritis of the hepatic artery and aneurysmal thrombosis and vasculitis of the superior mesenteric artery leading to intestinal infarction have been reported [44]. Aneurysms are usually seen in the saccular form and show curvilinear, thickened walls, with partial thrombosis on CT or MR angiography. Aortic aneurysms can be encountered in multiple levels and may involve the orifice of visceral arteries (Figures 22).



Figure 21: A 21-year-old male patient with both chronic and acute stage thrombosis of portal venous system in Behcet's disease. A. Axial CT scan shows chronic thrombosis as filling defect in the lumen of small-calibered main portal vein (arrowhead) and it is seen as cavernous transformation with multiple tortuous vascular channels in the porta hepatis (arrow). B. Acute stage thrombosis is seen as thrombosis completely filling and expanding portal vein confluence and splenic vein without collateral formation.

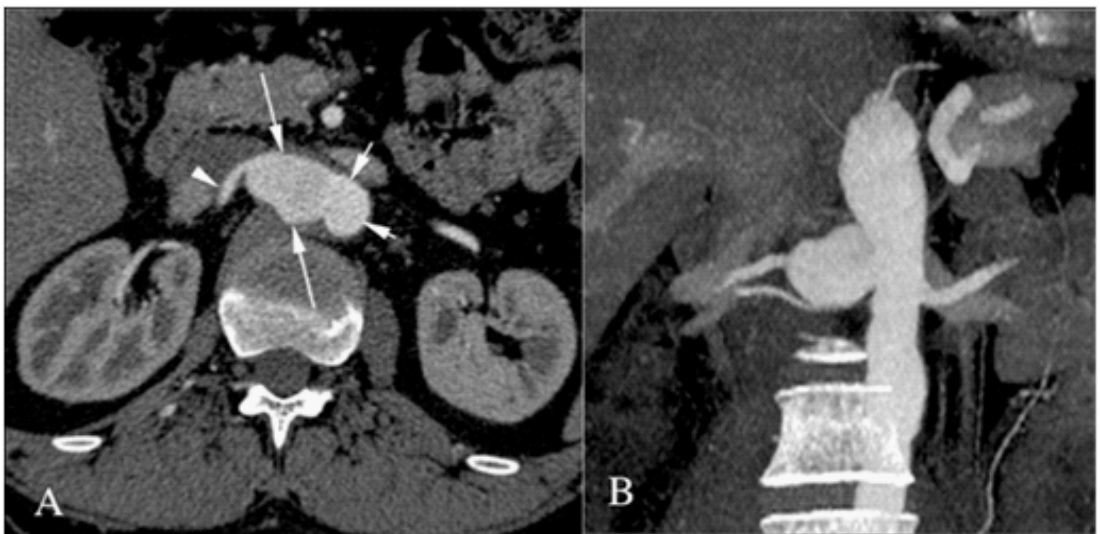


Figure 22: A. Axial and B. Coronal reformatted CT images of the same patient in figure 8 show saccular aneurysm (long arrows) originated from right side of the abdominal aorta (short arrows) and extended toward right renal artery orifice (arrowhead).

Invasive procedures, whether diagnostic or therapeutic, are not recommended for Behcet's disease patients with vascular involvement, because the development of recurrent false aneurysms at anastomotic or traumatic sites such as the angiographic puncture site is an important concern in Behcet's disease patients. Thrombosis formation is also frequent following venous intervention. CT and MR angiography are safer than conventional angiography for analyzing vascular involvement in these conditions [41,63].

Extra-Intestinal Organ Involvement

Pancreas

Pancreatic involvement was seen in 2.9% of cases in an autopsy series from Japan [12]. Acute pancreatitis due to Behcet's disease is rarely reported [64]. Pancreatic inflammation is likely due to vasculitis [58].

Liver

Budd-Chiari syndrome is the most common manifestation of liver involvement in patients with Behcet's disease as described above in the section on venous involvement. In addition to Budd-Chiari syndrome, hepatomegaly can be seen due to fatty liver and congestion. Acute and chronic hepatitis, cholelithiasis and cholecystitis, primary biliary cirrhosis, hepatic abscesses, and sclerosing cholangitis [12,65-67] are also-but rarely-seen.

Spleen

Splenic involvement was reported in 37 of 170 autopsies in Japan, including splenitis, splenomegaly, hemosiderosis, infarction, and auto-splenectomy [12]. A slight enlargement of the spleen unrelated to portal hypertension has been observed in 20% of male patients [68].

Kidney

Because Behcet's disease affects all types and sizes of blood vessels and vascular thrombosis is seen in one third of cases, a wide spectrum of renal lesions can be encountered in Behcet's disease [69]. Characteristic renal lesions in Behcet's disease are secondary amyloidosis, glomerulonephritis, renal vascular disease, and interstitial nephritis. Behcet's disease patients may present with different clinical conditions ranging from asymptomatic hematuria and/or proteinuria to end-stage renal disease [69]. US can show non-specific renal morphological changes. Doppler US can evaluate vascular involvement such as stenosis, occlusion, and aneurysm formation. Obesity and artifacts caused by air in the intestines can make it impossible to examine vascular structures. In addition, mild arterial stenosis and small aneurysms can be missed by Doppler US. As a result, CT and MR angiography have higher efficacy in the evaluation of vascular lesions.

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