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UNUSUAL CAUSES OF RIGHT ILIAC FOSSA PAIN – RADIOLOGICAL FEATURES WITH HISTOPATHOLOGICAL CORRELATION.

Radiology

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ABSTRACT

Pain in the right iliac fossa is a common clinical scenario and can result from a variety of causes, the most common being acute appendicitis. Other common causes of right iliac pain include, ureteric calculi, intussusception, ectopic pregnancy, cecal neoplasm, tubo-ovarian pathology and ileocaecal tuberculosis. Beyond these, a number of rarer conditions can cause pain in the iliac fossa. This article illustrates the radiological findings of some of these rarer causes of right iliac fossa pain, correlating them with the histopathological features.

KEYWORDS

Right iliac fossa, computed tomography, histopathology

Introduction

Right iliac fossa pain requires prompt investigation and diagnosis. While acute appendicitis is the most common etiology, the cause is not always obvious so other differential diagnoses must be considered.[1] Diagnostic imaging plays a key role in diagnosis, and can help to determine proper management. Our goal is to offer insight into some of the rarer causes of right iliac fossa pain, highlighting their characteristic imaging appearances with histopathological correlation

DESMOID TUMOR

Desmoid tumors result from mutations involving fibroblasts, causing uncontrolled proliferation. They are quite rare, with 2-4 cases per million persons per year.[2] Desmoid tumors can present sporadically, or they can be associated with inherited syndromes such as familial adenomatous polyposis Gardner syndrome.[3]They constitute about 3% of all soft tissue tumors[4], occurring most commonly in women in their mid-30s.

Growth rates are variable, and they do not metastasize.[5] Abdominal desmoids can occur in the abdominal wall, mesentery, or retroperitoneum.[6] In addition to the mentioned inherited syndromes, risk factors include a history of abdominal or pelvic surgery, trauma, pregnancy and estrogen therapy.[6]

Desmoid tumors exhibit a high rate of local recurrence (20–77%) depending on the location, extent and completeness of the initial resection. Computed tomography (CT) and magnetic resonance imaging (MRI) features vary depending on the composition. Abdominal desmoid tumors usually present as a soft tissue attenuation value lesion in the rectus abdominis muscle and the adjacent aponeurosis. Typically, they are well circumscribed, but aggressive lesions can show ill-defined margins. Compared to muscle, the can be hypo, iso or hyperdense. Contrast enhancement will be variable.[6] Histopathological analysis should be done for definitive diagnosis.[5]

Imaging

A 60-year-old man presented with lower abdominal pain and swelling in the right iliac region for one month. Based on the contrast enhanced CT (CECT) findings (Fig. 1A, 1B), a desmoid tumorwas suspected and the patient was referred to surgery.

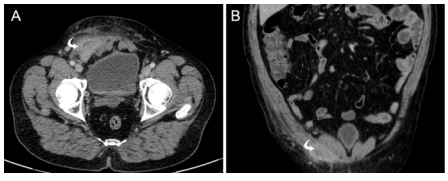

Figure. 1 A,B. A 60-year-old man presented with lower abdominal pain and swelling in the right iliac region for one month. Post contrast images of the abdomen in axial view (A) and reformatted coronal view in venous phase (B) showed a well-defined peripherally enhancing soft tissue lesion with perilesional fat stranding and central areas of necrosis (curved arrow) in the intramuscular plane of anterior abdominal wall in the right iliac fossa.

Histopathology

The patient underwent percutaneous needle biopsy, which revealed fibrocollagenous and adipose tissue with an infiltration of eosinophils (Fig. 1C, 1D and 1E). Edema and focal myxoid changes were noted. By immunohistochemistry, the tumor cells were focally positive for beta-catenin and diffusely positive for vimentin (Fig. 1D, 1E). ALK, CD34 and S100 stains were negative. The staining patterns were interpreted as favoring desmoid tumor.

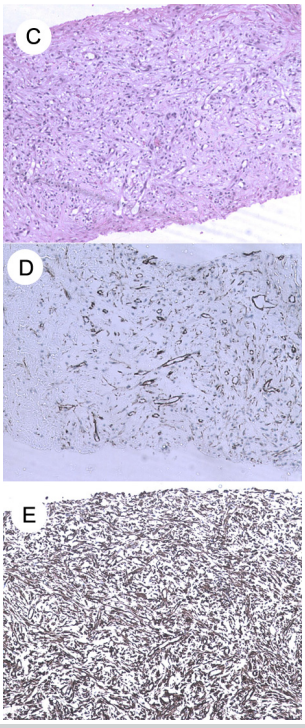

Figure. 1 C,D. Histologic section (C) (10x) showing fibrocollagenous and adipose tissue with an increased infiltration of eosinophils. Edema and focal myxoid change can also be seen. Imunohistochemistry showing lesional cells focally positive for beta-catenin(D)(10x) and diffusely positive for vimentin (E)(10x).

CAECAL LYMPHOMA

The ileocaecal region is a relatively rare site for involvement by non-Hodgkin's lymphoma, with an incidence of about 1.7%.[7][8]Diffuse large B cell type is the most common histology. Other types that can affect the colon include mantle cell, Burkitt's and Burkitt-like lymphoma. The most common gastrointestinal tract symptoms of NHL include abdominal pain (52%), hematemesis (23%, with mostly gastric involvement), obstruction (19%), vomiting (19%) and palpable mass (17%).[7]

CT is a primary tool for staging the disease, the usual appearance being a slightly or moderately enhancing, homogeneous soft tissue attenuation value mass. Diffuse infiltration of the bowel is usually seen with an intact mucosal surface. An intraluminal component will usually be minimal.[9]

It should be noted that the most common malignant lesion involving the ileocolic area is adenocarcinoma. The standard primary treatment of non-metastatic colon carcinoma is colectomy and lymph node dissection, followed by 5- flurouracil and oxaliplatin based chemotherapy.[10]Differentiating lymphoma from carcinoma before primary surgical management is crucial, because it plays a significant role in guiding management and influences both clinical outcome and quality of life.[11]

Imaging

A 51-year-old man presented with one month of right-sided abdominal pain and weight loss. CECT findings suggested metastatic caecal carcinoma (Fig. 2A, 2B).

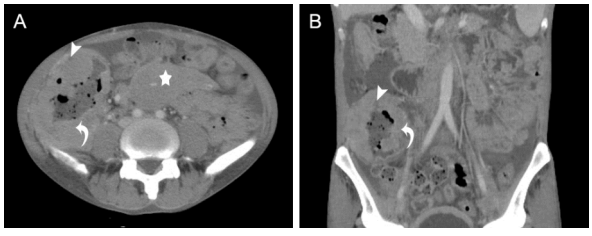

Fig. 2 A,B : A 51-year-old man presented with one month of right-sided abdominal pain and weight loss. Post contrast axial (A) and reformatted coronal image (B) in venous phase showing diffuse heterogeneously enhancing circumferential wall thickening (curved arrow) with areas of necrosis (arrow head) involving the caecum. Multiple heterogeneously enhancing lymph nodes are seen in the peritoneum, largest measures 4.4 x 7.6 cm in the aortic bifurcation (asterix).

Histopathology

Colonoscopy biopsy specimen showed colonic mucosa with extensive ulceration and atypical lymphoid cells in the lamina propria (Fig. 2C, 2D). The mitotic index was 11 per 10 high-power fields (HPF). Increased apoptosis was also seen. Cellular features were highly suspicious for non-Hodgkin's lymphoma (NHL). Immunohist ochemistry (IHC) showed the atypical lymphoid cells to be positive for CD20, and scattered CD3 and vimentin positive cells were seen in the background. This confirmed the diagnosis of NHL, diffuse large B cell type.

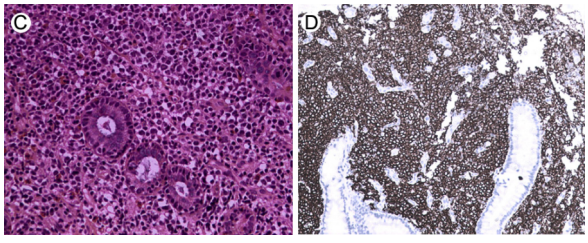

Fig. 2 C,D. Histologic section (C) (20x) shows colonic mucosa with extensive ulceration and increased number of atypical lymphoid cells in the lamina propria. IHC showing atypical lymphoid cells positive for CD20 (D)(10x).

SOLITARY CAECAL DIVERTICULITIS

Solitary caecal diverticulitis is a rare cause of acute abdominal pain and the etiology remains unclear. The incidence of a diverticulum involving the caecum is rare, reportedly 0.04% to 2.1%.(12,13)

The CT findings of caecal diverticulitis are similar to those of left sided diverticulitis, which includes focal pericolonic inflammation, colonic wall thickening, diverticula, thickening of the adjacent fascia, and extra luminal mass effect.

A study by Chou et al proved the accuracy of ultrasound in diagnosing caecal diverticulitis with a sensitivity of 91.3% and a specificity of 99.5% in differentiating right-sided diverticulitis from appendicitis. In another study,CT scanning can improve the pre-operative diagnosis to rule out malignancy.(14,15)However most of the cases of caecal diverticulitis are diagnosed during surgery for presumed caecal malignancy or appendicitis and its complications.

Imaging

A 70-year-old woman presented with right-sided abdominal pain. Ultrasound of the abdomen demonstrated a relatively well-defined, predominantly hypoechoic lesion in the right iliac fossa.(Fig.3A) No areas of vascularity were noted within. The appendix was not separately visualized. A CECT examination followed and was interpreted as suspicious for caecal malignancy.(Fig.3B, 3C) A right hemicolectomy was performed, Surgery revealed an exophytic lesion measuring 4 cm, arising 1cm distal to the ileocaecal junction. A right hemicolectomy was performed.

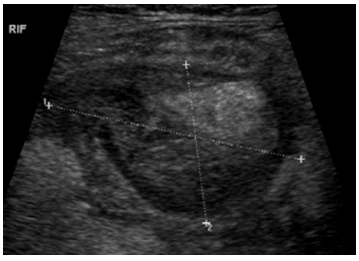

Fig. 3 A. A 70-year-old woman presented with right-sided abdomen pain. Ultrasound of the abdomen showing a relatively well-defined mixed echoic lesion, predominantly hypoechoic of size 5.5 x 5 cm in the right iliac fossa.

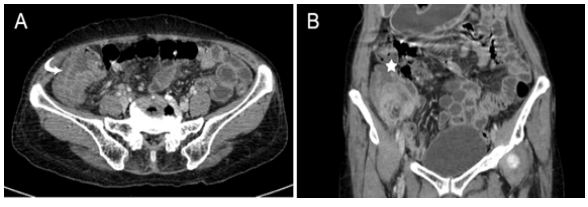

Fig. 3B,C. A 70-year-old woman presented with right-sided abdomen pain. Post contrast axial (B) and reformated coronal view (C) of CECT of the abdomen in venous phase showed an ill-defined heterogeneously enhancing mass involving the cecum (curved arrow). Appendix was not seen separately. Focal loss of fat plane with the terminal ileum and ileocaecal junction was seen (asterix).

Histopathology

On microscopy, the specimen showed diverticulitis with focal abscess formation (Fig. 3D, 3E). The serosa was involved by the inflammatory process. There was no evidence of inflammatory bowel diseases, tuberculosis or malignancy.

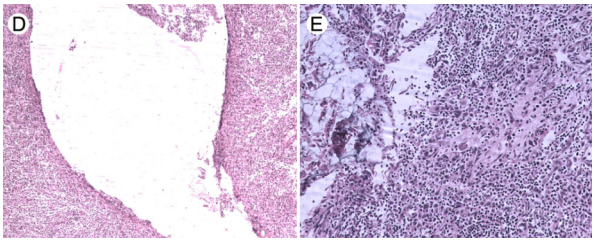

Fig. 3 D,E. Histologic section (D) (4x) and (E)(10x) showing diverticulitis with focal abscess formation.

GASTROINTESTINAL STROMAL TUMOR OF THE ILEUM.

Gastrointestinal stromal tumors (GISTs) account for less than 1% of gastrointestinal tumors, yet are the most common mesenchymal tumors of the GI tract.[16] GISTs are commonly seen in the stomach, followed by the small intestine. They are distinguished by the characteristic KIT or platelet-derived growth factor receptor alpha (PDGFRA) gene mutations. The most common age of presentation is between 50 and 80 years of age and there is no sex predilection.[17] The tumors generally occur sporadically, but rare familial forms are also observed. Metastasis occurs in approximately 10% to 25% of patients.[18]Gastrointestinal bleeding (40%) is the usual mode of presentation, followed by abdominal mass (40%) and abdominal pain (20%). The risk of GIST progression is assessed by mitotic index, tumor size and tumor location.[19]

CT imaging is the initial modality of choice if a GIST is suspected.CT characteristics of GISTs vary depending on the risk stratificatio n.[20][21]Ileal GISTs can be intraluminal, mural or extraserosal. Commonly they are heterogeneously enhancing but a homogeneous pattern does occur less frequently. Areas of low attenuation within the tumor can represent from cyst formation, hemorrhage or necrosis.[21]

Imaging

A 51-year-old man presented with four months of diffuse abdominal pain, prompting a CECT (Fig. 4A, 4B) The two probable radiological diagnoses included GIST arising from the distal ileum or peritoneal sarcoma with bowel wall infiltration. The patient underwent an exploratory laparotomy.

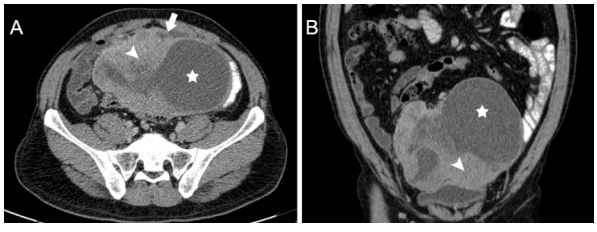

Fig. 4A,B. A 51-year-old man presented with four months of diffuse abdominal pain. Post contrast axial (A) images and reformatted coronal images (B) in venous phase with oral contrast show a large ovoid heterogeneously enhancing mass lesion with both solid (arrow head) and cystic components(asterix) in the peritoneal cavity occupying the lower abdomen. The fat plane between the lesion and the distal ileum is lost (down arrow).

Histopathology

Upon bowel dissection, the lesion seemed to arise from the submucosa. Focal necrotic areas were present within. On microscopic examination, there were spindle-shaped cells with a mitotic index of one per 50 HPF (Fig. 4C). The tumor cells were strongly positive for vimentin, DOG1 and CD117 (Fig 4D) and negative for SMA and S100. Features were consistent with a high risk, grade I GIST (spindle cell type).

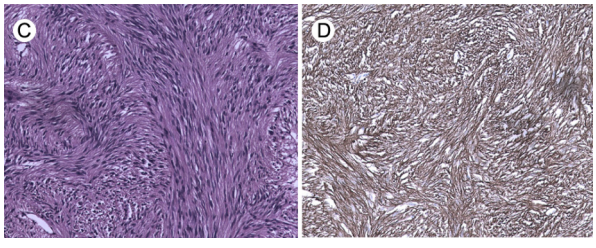

Fig. 4 C,D. Histologic section (C) (10x) shows spindle-shaped cells. The tumor cells showing strong positivity for CD117 (D)(10x).

EXTRA-ADRENAL PARAGANGLIOMA

Extra-adrenal retroperitoneal paragangliomas are rare tumors with an incidence of 2-8 per million. These tumors arise from embryonic neural crest cells and are composed mainly of chromaffin cells.[22]They can occur at any site, but most commonly in close relation to the sympathetic nervous system and aorta.[23]They are commonly seen during the 4th to 5th decades of life and show no sex predilection. [24] Retroperitoneal paraganglia are symmetrically distributed along the abdominal aorta and are closely related to the sympathetic nervous system. They can be functioning or non-functioning. The functioning type synthesizes, stores and secretes catecholamines and thus produce symptoms such as palpitations, headaches, sweating and symptoms of hypertension. Non-functioning ones can remain clinically silent with vague symptoms.[24] Extra-adrenal retroperitoneal paraganglioma causes a dilemma in both diagnosis and treatment. On CT, smaller tumors usually appear homogenous with sharp margins, whereas larger ones usually are heterogeneous. They typically show intense enhancement with contrast administration.[24]

Imaging

A 55-year-old man presented with pain in the right iliac fossa. The CECT features were suggestive of paraganglioma (Fig. 5A, 5B). A laparotomy and excision of the lesion followed.

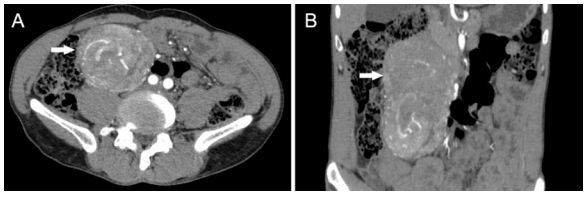

Fig. 5A,B. A 55-year-old man presented with pain in the right iliac fossa. Post contrast axial view (A) and reformatted coronal view (B) of the abdomen in arterial phase show a well-defined heterogeneously enhancing mass lesion (right arrow) involving the right para-aortic region.

Histopathology

On microscopy, the lesion was encapsulated with tumor cells arranged in a predominantly nested pattern. Focal areas showed tumor cells in sheets, interspersed by abundant blood vessels (Fig. 5C, 5D). Individual tumor cells showed abundant eosinophilic cytoplasm and pleomorphic nuclei with few showing hyperchromasia. The mitotic index was 20 per 10 HPF. Focal areas of necrosis also were seen. The pathological features confirmed the diagnosis of an extra-adrenal paraganglioma

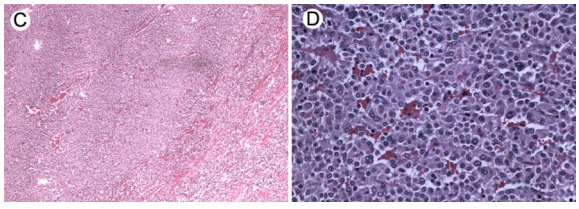

Fig. 5C,D. Histologic section (C) (4x) and (D)(20x) showing tumor cells arranged predominantly nested pattern and focal areas showing tumor cells in sheets interspersed by abundant blood vessels. Individual tumor cells showed abundant eosinophilia cytoplasm and pleomorphic nuclei with few showing hyperchromasia.

RETROPERITONEAL LEIOMYOSARCOMA.

Retroperitoneal leiomyosarcomas account for 13% of all adult soft-tissue sarcomas.[25] The most common age of presentation is 50 to 70 years with a female predilection. The usual presenting symptoms are due to mass effect or local invasion.[26] For non-metastatic tumors, the local extent is an important factor in determining the optimal management options and prognosis.[27] CT scanning in particular is useful for localization, as well as predicting local and remote spread.[26] Retroperitoneal leiomyosarcoma is typically seen on CT as a solid non-fatty mass with large conspicuous zones corresponding to areas of necrosis.[28]

Imaging

A 46-year-old woman who is following up after a hysterectomy presented with right iliac fossa pain. A CECT was performed (Fig 6A, 6B). Along with the abdominal findings, the lung bases revealed multiple nodules. A retroperitoneal tumor was considered to be the most likely diagnosis. A CT guided biopsy was performed on a lung nodule, which proved to be high-grade leiomyosarcoma.

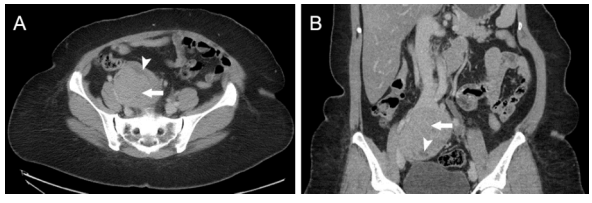

Fig. 6 A,B. A 46-year-old woman who is following up after a

hysterectomy presented with right iliac fossa pain. Post contrast axial (A) and reformatted coronal view (B) of the CECT of abdomen in venous phase shows well defined heterogeneously enhancing mass lesion (left arrow) with necrosis/cystic changes (arrow head) in the retroperitoneum at the confluence of the bilateral common iliac veins into the inferior venae cava.

Histopathology

A core biopsy showed hypercellular spindle cells lesions with moderate nuclear atypia (Fig. 6C, 6D). Mitosis was 12 per 10 HPF. No evidence of necrosis was seen. Because of atypia and increased mitoses, a diagnosis of leiomyosarcoma was rendered.

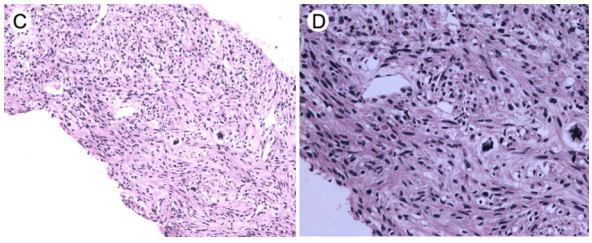

Fig. 6C,D. Histologic section (C) (10x) and (D)(20x) showing hyper cellular spindle cells lesion with moderatenuclear atypia. A few atypical mitoses can also identified

PARASITIC LEIOMYOMA.

Parasitic leiomyomas ("wandering fibroids") are rare benign tumors.[29] The etiopathogenesis is not well understood, but could be related to separation of the lesion from the uterus, with establishment of blood supply from adjacent structures such as the bowel, peritoneum, omentum or mesentery.[30] Most parasitic leiomyomas occur after laparoscopic myomectomy, presumably arising from leiomyoma fragments left behind.[31] Occasionally a pedunculated subserosal leiomyoma can twist on its pedicle, and can become free in the peritoneal cavity. The separated lesion can adhere to surrounding structures and can cause complications including intestinal obstruction and pain. [32]

The most reliable imaging modality for diagnosis is MRI. A typical leiomyoma will be T1 isointense and T2 hypointense. On CT scan the mass will mostly be homogeneous with variable enhancement on contrast administration. But the lesion can also be heterogeneous depending on the extent of degeneration, fibrosis, and calcification. Ultrasonography will show a whorled appearance, with variable echogenicity.[33]

Imaging

A 43-year-old woman presented with vague abdominal pain and weight gain. A possibility of a wandering fibroid was made via CECT.(Fig. 7A, 7B). The patient underwent a laparotomy and excision of the lesion.

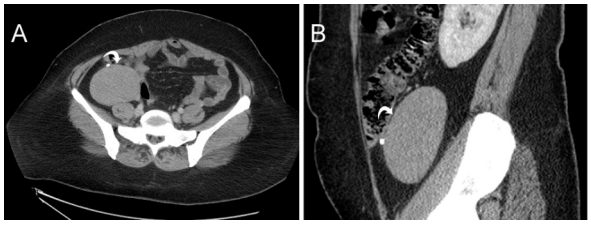

Fig. 7 A,B. A 43-year-old woman presented with vague abdominal pain and weight gain. Post contrast axial view (A) and reformatted sagittal view (B)of CT abdomen in venous phase showed well-defined homogenously enhancing mass lesion (curved arrow) in the right iliac fossa. A small metallic clip noted anterior to the mass lesion. Similar metallic clips were also seen in the para uterine region. Based on these findings we considered the possibility of a wandering fibroid.

Histopathology

On microscopic examination, the lesion was as a spindle cell neoplasm with marked degenerative nuclear features. The mitotic count was 4 per 10 HPF. No necrosis was seen. By immunohistochemistry, the tumor cells were positive for vimentin and SMA and negative for S100. The Ki67 index was 5 per 10 HPF.(Fig 7C, 7D, 7E)These features were consistent with a diagnosis of symplastic leiomyoma, which was in accordance with the radiological diagnosis.

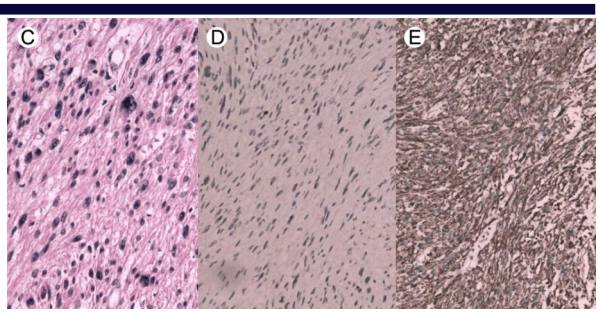

Fig 7 C,D,E. Histologic section (C) (10x) showing spindle cell neoplasm with marked degenerative nuclear features. The tumor cells show positive staining for vimentin (D) and SMA (E).

CONCLUSION

Radiological and pathological features of right iliac fossa masses with seven rare diagnoses are discussed. A high index of suspicion and knowledge of lesions other than appendicitis and colon carcinoma will help to establish a correct diagnosis, sometimes with a profound impact on management and prognosis.

COMPLIANCE WITH ETHICAL STANDARDS

Ethical approval

All procedures in the studies involving human participants were performed in accordance with the ethical standards of the institutional and/or national research committee.

Informed consent

Informed consent was obtained from all individual participants included in the study. Consent was obtained for publishing the case and the radiological images. No information has been included in the article that would reveal the identities of the patients.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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