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Case Report

A case of bilateral posterior mediastinal myelolipoma ☆

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ABSTRACT

A 71-year-old female presented with chronic shortness of breath and underwent routine examination at the emergency department. A plain chest radiograph revealed a large lobulated posterior mediastinal mass that was incidentally found to be unrelated to the main complaint. Further cross-sectional images were obtained to characterize the lesion, which revealed bilateral involvement of a prevertebral mixed attenuation large mass with minimal enhancement postcontrast administration. Images were not conclusive in which the patient underwent ultrasound-guided biopsy and further histopathological examination, which revealed a myelolipoma of the posterior mediastinum, a rare entity to be seen at that location. Here, we present the case of posterior mediastinal myelolipoma.

Introduction

Myelolipoma is a rare benign extra-adrenal lipomatous soft tissue neoplasm. The occurrence of myelolipoma in an extra-adrenal site is rare. However, frequent locations of extra-adrenal myelolipoma are in the abdomen, pelvis, or retroperitoneum [1]. Myelolipoma was first reported by Edgar von Gierke in 1905 [2], and first named in 1929 [3]. The first posterior mediastinal myelolipoma (PMM) was described in English literature in 1984 [4]. The etiology of myelolipoma is unclear. According to the literature review, several hypotheses tried to explain the pathology [5]. PMM is usually discovered incidentally by imaging studies and confirmed by histopatho-

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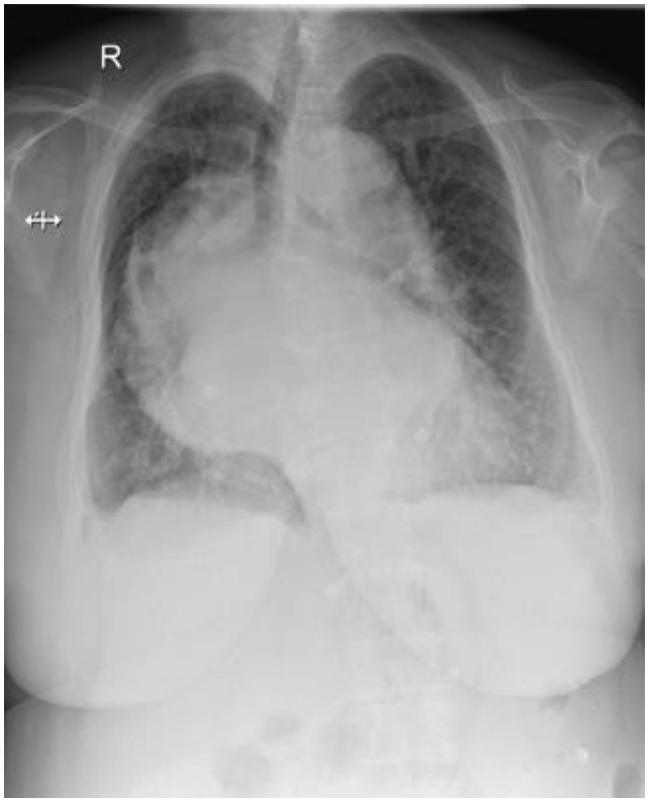

Fig. 1 – Frontal plain chest radiograph shows a large lobulated posterior mediastinal mass. Splaying of the carina and unfolded aortic arch are noted.

logical studies. It presents as a large, painless, and palpable mass mostly seen in the elderly. The mainstay of treatment for the mass is video-assisted thoracic surgery. The tumor was benign with no reported cases of recurrence or metastasis after surgical resection. Fewer than 30 cases of posterior mediastinal myelolipoma were discovered during a review of English literature. Accordingly, the presented case is an elderly female patient with an incidental posterior mediastinal mass that has been shown by histopathological studies.

Case report

A 71-year-old female presented to the emergency department by her family members with the main complaint of chronic shortness at rest, which lasted for 2 months and was aggravated by minor activity, which they thought to be coronavirus disease 2019 (COVID19) infection that was endemic at the time of presentation. The patient declared a history of generalized fatigability as well as left lower limb swelling, which was present for a long time. No other respiratory or abdominal symptoms were reported. The patient's past medical history included diabetes, chronic kidney disease (CKD) on hemodialysis, hypertension, and iron deficiency anemia. She was noncompliant with medications for these comorbidities. The patient did not seek medical assistance before. On physical ex-

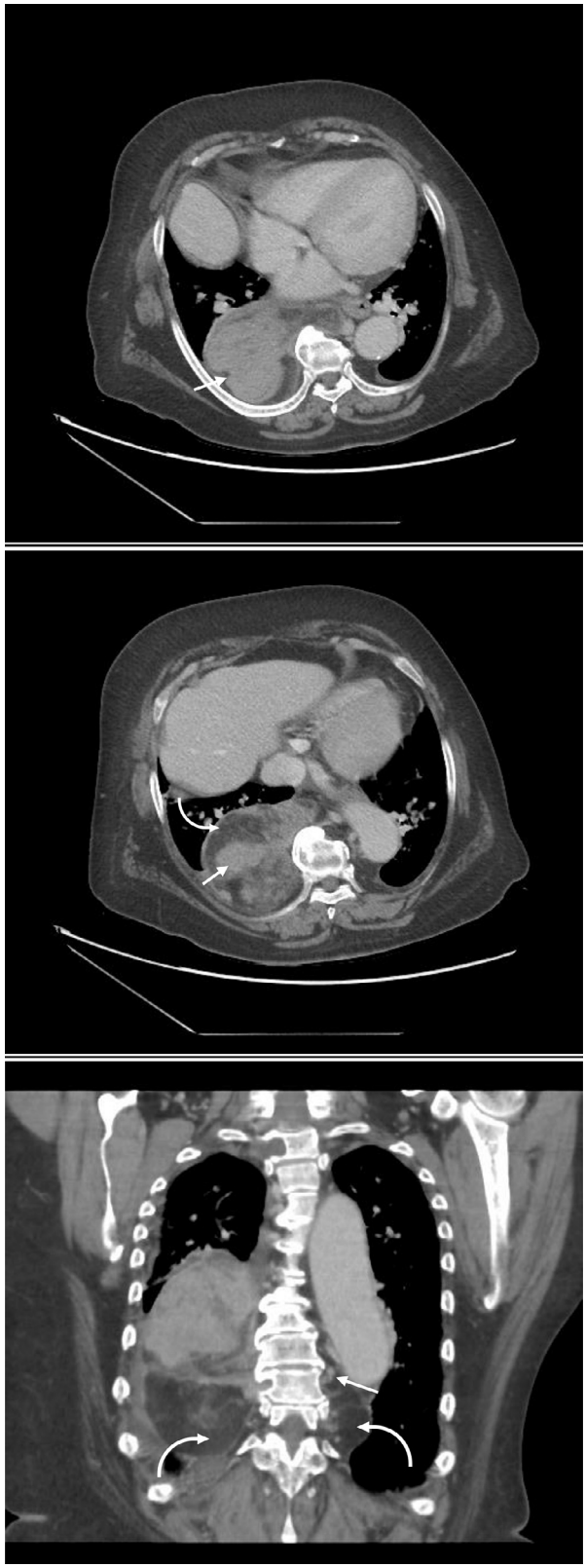

Fig. 2 – Three selected axial and coronal enhanced (CT) scans of the chest show a large mid-thoracic posterior mediastinal and/or paravertebral mass with mixed fat (curved white arrow) and soft tissue (straight white arrow) attenuation bilaterally.

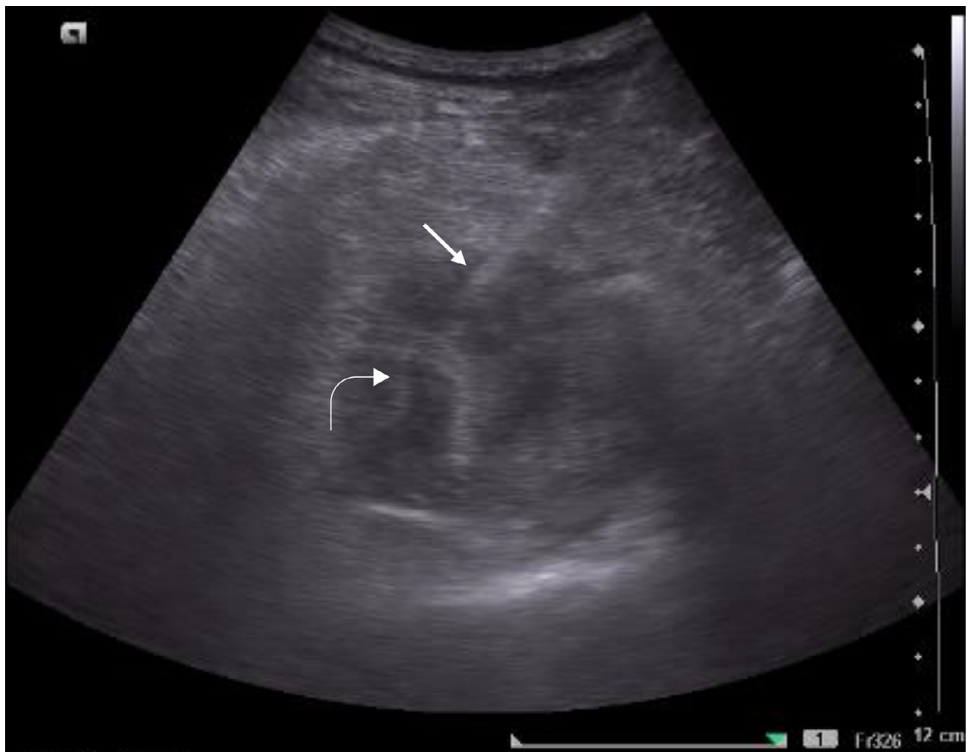

Fig. 3 – Ultrasound-guided biopsy showing the needle (straight white arrow) within the mass (curved white arrow) for tru-cut biopsy.

amination, the patient was ill-looking with palor skin. The respiratory examination revealed bilateral basal crepitations with no other findings. The abdomen was distended but without tenderness on palpation. The lower limbs showed bilateral pitting edema. The vital signs of this patient at the time of presentation showed a low-grade fever of 101.3 Fahrenheit (°F), the pulse was 110 beats per minute (BPM), systolic blood pressure was 113 millimeters of mercury (mm Hg), and diastolic blood pressure was 73 (mm Hg).

As per the hospital protocol, the patient was isolated to exclude the possibility of COVID-19 infection before continuing the investigations in which polymerase chain reaction (PCR) tests showed negative results. Echocardiography was normal.

A chest plain radiograph was obtained, which revealed a large lobulated heterogeneously opaque mass of the posterior mediastinum predominantly on the right side. There was splaying of the carina and unfolded aortic arch (Fig. 1). An enhanced computed tomography (CT) scan is advised to further characterize the lesion. Since the patient was diagnosed with CKD, dialysis was initiated after contrast administration. A nonenhanced CT scan of the chest was performed because of patient renal pathology, which revealed a large midthoracic posterior mediastinal and/or paravertebral mass with mixed fat, and soft tissue attenuation with a mean Hounsfield unit (HU) of 34 of the soft tissue components. It measured 8.2 × 7.6 × 12.7 cm in the transverse, anteroposterior, and craniocaudal dimensions, respectively. After patient consent, an enhanced CT scan of the chest was performed for further elaboration. On enhanced CT scan, the mass revealed homogenous enhancement with a mean HU of 95 of the soft tissue component (Fig. 2). There was encasement and displacement of the lower portion of the azygos vein and abussion of the right main pulmonary trunk. However, the bronchial tree showed no bilateral narrowing, and the lungs had subsegmental atelectasis. There was no evidence of bony erosion in the nearby rib or vertebra, no neural foraminal narrowing, no pleural effusion or surrounding tissue invasion, and no calcifications identified. Differential diagnoses were suggested to be myelolipoma, liposarcoma, thymolipoma, and germ cell tumors. The diagnostic plan was to perform an ultrasonographic assessment of vascularity along with a tissue biopsy. On ultrasound (US)-guided biopsy, there was no intense vascularity of most of the mass. Under the aseptic technique, the patient was in a lateral decubitus position since the patient could not tolerate being in a prone position, an 18-gauge tru-cut biopsy needle was inserted through a right posterior intercostal approach, and biopsy was taken from the hyperechoic component with no post biopsy complications (Fig. 3).

Histopathological examination revealed predominant mature adipose tissue with interspersed islands of trilinear hematopoietic cells, including megakaryocytes without any signs of atypia or malignant transformation, consistent with the diagnosis of myelolipoma.

A microscopic examination revealed predominant mature adipose and hematopoietic tissue with intermingling (Fig. 4,5).

Ultimately, it was concluded that the patient's main complaint was due to chronic renal disease rather than the mass. The management plan was to optimize the patient clinical condition and keep the patient on periodic imaging follow-up for the mass to look for interval growth or mass effect on vital

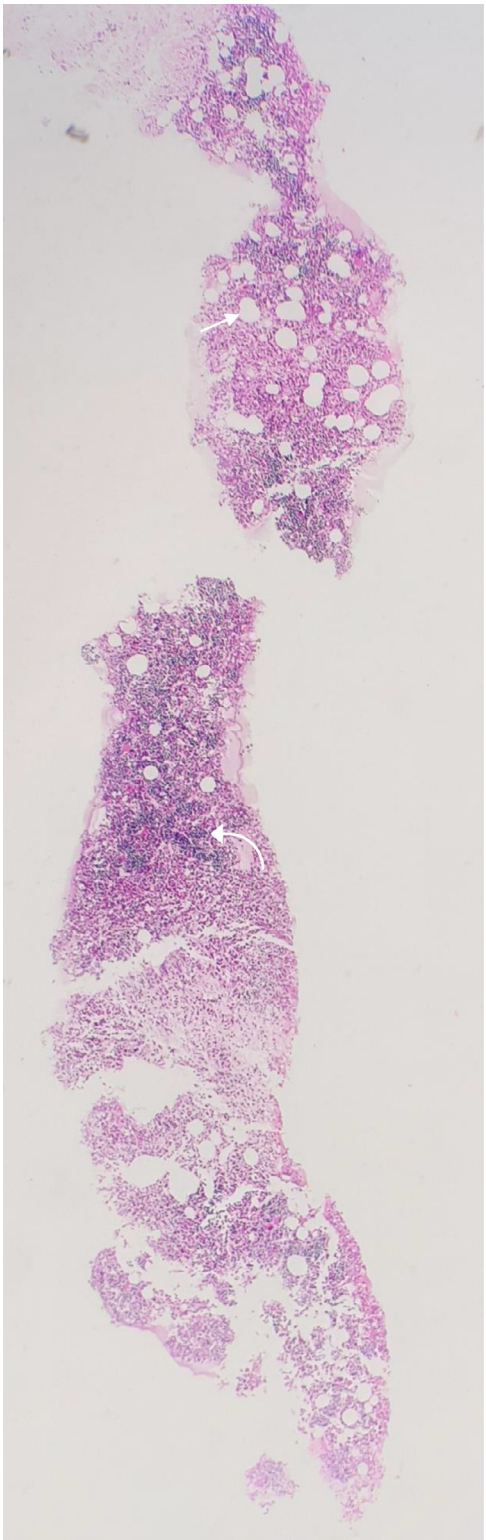

Fig. 4 – Low-power photomicrograph at 4X: Core of hematopoietic elements (white curved arrow) with intermixed mature adipocytes (white straight arrow).

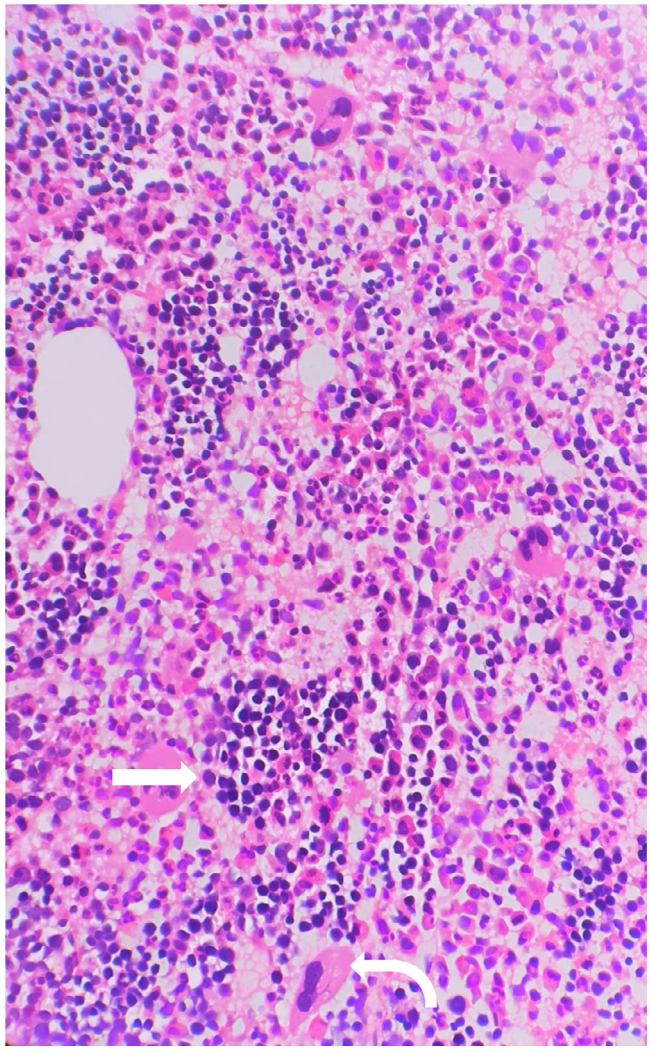

Fig. 5 – High-power photomicrograph at 40X: Trilineage hematopoietic elements (white straight arrow), including scattered megakaryocytes (white curved arrow).

structures because the patient was not a surgical candidate for intervention due to uncontrolled comorbidities. The patient was discharged after optimization of comorbidities and scheduled for a follow-up clinic.

Discussion

Myelolipoma was first reported by the pathologist Edgar von Gierke in 1905 [2] and first named in 1929 [3]. The first PMM was described in English literature in 1984 [4]. Myelolipoma is a rare benign tumor consisting of 2 types of tissue: mature adipose tissue and hematopoietic tissue [1]. Three to 5% of myelolipoma was found to be on the adrenal glands. Extra-adrenal myelolipomas are rare and include the presacral region, stomach, liver retroperitoneum, spleen, mediastinum, and lungs, where the mediastinum is approximately 3% of all myelolipomas, mostly in the right lower posterior medi-

astinum. There have been no reports of myelolipomas transforming into malignant tumors [6]. Less than 39 cases of mediastinal myelolipoma have been reported to date according to the literature done in 2018 [1,5].

The etiology of myelolipoma is not clearly understood according to the literature. Several hypotheses have tried to explain the pathophysiology, which includes 4 hypotheses. The first hypothesis is the most likely hypothesis, which describes myelolipoma as an ectopic adrenal or hematopoietic stem cell affected by triggering factors such as trauma history, obesity, hypertension, chronic inflammation, malignant tumors, or endocrine disorders. The second hypothesis described it as an evolving cell from a metaplastic process of embryonic primitive mesenchymal cells or embolism of bone marrow cells that travels through the bloodstream. Furthermore, a third hypothesis described it as a mutation in the chromosome leading to a neoplastic phenomenon. The last hypothesis described it as an ectopic hematopoietic tissue that may contain stem cells where myelolipoma would originate because some cases were related to hematological disorders [5].

Moreover, most of the patients with mediastinal myelolipoma presented in their sixth decade, and there was no significant relationship between the sex of the patients, and the tumor [7,8]. Diagnosis is made incidentally on visits for routine clinical examinations with no or unrelated clinical symptoms, as presented in this case. However, some patients with a large mass that causes a mass effect on the adjacent anatomic structures, which results in a variety of symptoms, such as productive cough, stiff neck, and dull back pain [7,8].

Physical examination and blood tests have no significant value on the diagnosis, while chest X-ray, CT scans, and magnetic resonance imaging (MRI) are helpful to the diagnosis. The diagnosis is suggested on scan according to the margins of the lesion if it is a well-circumscribed mass and shows mixed densities. However, due to the lack of typical manifestations of myelolipoma, this leads to difficulty in definitive diagnosis without histopathological studies [7]. Therefore, no specific presentations can lead to a diagnosis of PMM, which should be differentiated from other similar tumors, such as neurogenic tumors, lymphomas, malignant mesotheliomas, and extramedullary hematopoietic tissue [9]. Moreover, one study described that the density of the tumor was approximately equal to the adipose CT value of 20-50 HU, including fat (−120 to −20 HU), and bone marrow measures (15-40 HU). In the case of contrast administration, no intense pattern of enhancement was observed. Furthermore, calcification occurrence within the tumor is rare, but it has been reported in the literature [10].

On the other hand, MRI showed isointensity or slight hyperintensity in T1- and T2-weighted images according to the dominant content of that mass, and after the administration of intravenous contrast media, the myeloid elements showed moderate enhancement, and adipose tissue showed no enhancement, resulting in heterogeneous enhancement [8].

The definite diagnostic test of myelolipoma is based on tissue biopsy and histopathological examination [11,12]. Microscopically, the masses are round, well defined, and encapsulated. The capsule measures approximately 4-5 centimeters (cm) in diameter [13]. Additionally, the manifestation of mediastinal myelolipoma is the same as myelolipoma in other locations, which consist of mature adipose tissue and hematopoietic cells, including myeloid, erythroid, and megakaryocytic elements, and the accumulation of lymphocytes is often present [14,15]. The presence of megakaryocytes is one of the essential elements for a definite diagnosis [16]. Myelolipoma might be associated with extramedullary hematopoietic tissue to differentiate between them histologically; the latter does not contain fat and is usually multifocal [17].

There has been controversy regarding the diagnostic and therapeutic algorithms of PMM. One study proposed that biopsy is not necessarily preoperative due to risks of hemorrhage and pneumothorax when surgery is deemed necessary [9]. However, the preparable method according to the literature is CT-guided needle biopsy, especially in asymptomatic patients, to avoid surgery; however, there is a risk of dissemination in cases of malignant neoplasms [18]. On the other hand, there has been a proposal that this mass should be resected if it grows more than 10 centimeters in diameter or if it is found to be small at the time of diagnosis due to observations of potential progressive enlargement, which results in increased risk to be adjacent to vital structures and hence potential surgical complications are increased, putting into consideration that most of the patients are present in old age [9].

The treatment options for primary mediastinal myelolipoma either by conventional thoracotomy or a video-assisted method depend on many factors, such as the size of the lesion or the development of local symptoms [7,9]. The mainstay of treatment for the mass is video-assisted thoracic surgery (VATS) [9]. There are many ways to perform VATS procedures, and the commonly used method is triportal VATS. Another approach is uniportal VATS; one case was performed using this technique, and the patient's position was adjusted from lateral to semiprone to facilitate the procedure through one access [9]. This procedure is related to many advantages, as it is minimally invasive compared with open thoracotomy, there is less trauma, and faster recovery. The characteristics of PMM during operations are a tumor with rich blood supplies that mainly arise from branches of the aorta or intercostal arteries, which makes it difficult to safely divide these vessels via VATS during resection of the mass to prevent excessive bleeding. One case reported bothersome bleeding due to an intercostal artery injury [9].

A study showed good outcomes regarding operation and minimal blood loss. Additionally, PMMs measured as 8 cm or less were proposed to be successfully resected via VATS. Regarding tumors of a larger size, open thoracotomy is more suitable as it might be associated with severe adhesions, which may result in excessive intraoperative results [9].

Conclusion

PMM is an extra-adrenal tumor of benign origin but can be bothersome if it is large enough to cause symptoms to the patient as well as if it is adjacent to vital structures. Most patients present asymptomatic with incidental PMM at old age. It can progressively enlarge with time.

Most authors have controversy regarding the diagnostic approach. There are no typical features on imaging. However, CT scans, and MRI are widely accepted noninvasive investigations to suggest the diagnosis. A confirmatory method of diagnosis is tissue biopsy with microscopic examination, but it can result in dissemination if malignancy is suspected, and a risk of bleeding in cases of rich blood supply.

Surgery is considered in cases of progressively enlarging tumors reaching more than 8 cm or symptoms resulting from the mass effect. VATS is a widely accepted therapeutic method due to certain advantages with minimal complications. No risk of recurrence of metastasis or malignant transformation was reported.

Patient consent

Informed written consent was obtained from the patient for publication of the case report and all imaging studies. Consent form on record.

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