Retroperitoneal mesenchymal chondrosarcoma with metastasis to iliac vein: A rare case report and review of the literature

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Abstract

The iliac vein is an extremely rare site of metastasis for extraskeletal mesenchymal chondrosarcoma (ESMC). Involvement of the veins usually leads to an extremely dismal prognosis. Here, we report a 50-year-old patient with retroperitoneal mesenchymal chondrosarcoma and initial metastasis to the iliac bone, which further progressed to involve the iliac vein. In the current study, we reviewed the major characteristics of ESMC and the previously reported cases considering the rarity of these tumors.

Keywords : Extraskeletal mesenchymal chondrosarcoma, iliac vein, metastasis, magnetic resonance imaging

Introduction

Chondrosarcoma (CS) is a heterogeneous type of neoplasm which is specified by cartilaginous matrix production through the tumor cells. CS is the third most prevalent bone malignancy after myeloma and osteosarcoma[1]. Different types of CS have been pathologically identified, such as mesenchymal, conventional, dedifferentiated, myxoid, and clear cell [2]. Mesenchymal chondrosarcoma (MCS) is a high-grade type of CS that was evaluated for the first time in 1959 by Bernstein and Lichtenstein in 1959[3]. The MCS is a rare and aggressive type of conventional chondrosarcoma that accounts for nearly 1% of all CSs and has a poor prognosis of 45–55% survival in 5 years [4]. 30% to 50% of MCSs have an extraskeletal origin. [5]. Most cases are diagnosed in the second decade of human life without any significant sex predilection [6]. In this case report, we present a case of ESMC with metastasis to the left iliac vein, which to the best of our knowledge, has not been reported before.

Case presentation

A 50-year-old woman with previously diagnosed retroperitoneal mesenchymal chondrosarcoma was referred to our hospital with the chief complaint of left leg pain and swelling. The pain started four months ago and increased gradually until the administration day. The pain was localized in the lumbosacral and radiated through the lateral part of her left lower limb, and it was not reduced by either rest or consumption of any analgesics. Her limb edema started one week before her admission, and she lost her ability to walk on her side. She did not smoke, consume alcohol, or illicit drugs and underwent treatment with chemotherapy. Her past drug history involved the consumption of daily warfarin(due to prior history of deep vein thrombosis) and opioid analgesics. On the physical examination, no ulceration or pigmentation was observed. No lymphadenopathy was found in the inguinal region. She has tender 4+ non-pitting edema on her left leg in conjunction with tactile tenderness of her lumbosacral region. Her right leg had limitations in mobility and movement. The ankle-brachial index was 1.02 in the left leg and 1.04 in the right leg.

The Computed Tomography (CT) images (Fig 1) displayed a minimally enhancing heterogeneous retroperitoneal mass with extensive dense popcorn-like calcifications located in the left paracervical area extending to the ipsilateral distended external, internal and common iliac veins

Magnetic resonance imaging (MRI) with and without gadolinium contrast media (Fig 2) revealed a large lobulated hetrosignal lesion with internal popcorn signal voids(calcifications) in the left paracervical area invading to proximal of the left external/internal iliac veins and the distal portion of the left common iliac vein. Both MRI and bone scan (Fig 3) demonstrate multiple bone metastasis in pelvic and both femoral bones,with largest deposit in right iliac wing,breaking through internal and external cortices and expanding subperiosteally with aggressive and massive periosteal reaction According to the MRI report, the tumor developed metastasis to the iliac vein, a rare location for mesenchymal chondrosarcoma to metastasis, and to the best of our knowledge, it has never been reported previously.

Discussion

Extraskeletal mesenchymal chondrosarcoma(ESMC) comprises a rare type of sarcoma originating from soft tissues, more frequently the meninges of the cranial and spinal cord, extremities, orbit and the lower extremities, particularly the thighs. Rarely, this tumor may arise from the retroperitoneum, kidneys, pancreas and hand musculature [7].ESMC account for about 1% of all chondrosarcomas[8]. Females are slightly more affected than males, while males carry a higher preponderance for extraskeletal and skeletal conventional chondrosarcomas[7]. ESMC has two peak ages of incidence: ESMC patients with involvement of the central nervous system, are generally younger (23.5 years old, range of 5–48 years old), while soft tissue and/or muscle tumor occur in older individuals (43.9 years old, range of 21-62 years old)[9]. Affected patients usually have unfavorable prognoses due to high rates of regional and distant metastasis[10].

Retroperitoneal mesenchymal chondrosarcoma has been reported rarely in the literature. In a cross-series study of Ghafoor et al. evaluating skeletal and extraskeletal mesenchymal chondrosarcoma imaging features, only 4 % of cases were located in the retroperitoneum [11]. ESMC can rarely involve the veins, with the cases being reported in the literature being as the primary site of the tumor. The iliac vein is an extremely uncommon site for ESMC and patients with primary iliac vein mesenchymal chondrosarcoma have a very

poor prognosis [12]. The femoral vein may also be the originating site for ESMC and it manifests with lower limb swelling and deep vein thrombosis[13]. Moreover, intravascular mesenchymal chondrosarcoma may also arise from the pulmonary veins[14].

ESMC displays a strong tendency to locally and distantly metastasize, which makes the clinical outcome extremely dismal, with a reported 10-year survival rate of 7-26 % [15]. In a study by Frezza et al. conducted on 113 patients with ESMC, Seventeen patients (15%) presented with distant metastasis at the time of diagnosis: seven patients (42%) had pulmonary metastasis, two patients(11%) showed bone metastasis and eight patients(47%) had metastasis to multiple organs[16]. Other previously reported sites of metastasis include lymph nodes[17] and the scalp[18], adrenal glands, pancreas and kidneys[11]. A review of the literature, revealed one case of retroperitoneal mesenchymal chondrosarcoma metastasizing to the vein. Juan Hu et al. presented a 61-year-old female with unintentional weight loss,persistent abdominal pain and nausea. Ultrasonography of the mass revealed two large retroperitoneal masses located adjacent to the inferior vena cava. The computed tomography scan showed dense and extensive, arc- and ring-like calcifications in the retroperitoneal soft tissue mass. Abdominal and pelvic magnetic resonance imaging(MRI) with gadolinium enhancement was also performed, which showed hypointensity on T1-weighted images and hyperintensity on T2-weighted images associated with peripheral speculated enhancement consistent with calcification. Subsequently, the histologic examination of the lesions revealed ESMC [9]. Our case is unique in that it was previously diagnosed with retroperitoneal mesenchymal chondrosarcoma with initial bone metastasis, which further progressed to involve the iliac vein.(Table 1)

MRI of ESMC often demonstrates equal or low signal intensity on T1W1 and heterogeneous hyperintense lesions on T2WI, as the intramural non-calcified and calcified regions of EMCS tend to have high and low intensity on T2WI, respectively, they are usually visible as areas of high signal intensity around areas with low signal intensity or the characteristic " salt and pepper" appearance[19]. Moreover, contrast-enhanced scanning of the lesions may show a diffuse nodular or heterogeneous pattern of enhancement, with the non-calcified component showing more homogenous enhancement and the calcified component showing less pronounced and heterogeneous enhancement, which can be clearly separated from each other in approximately 30 % of cases. Calcifications appear predominantly as chondroid type arc- and ring-like pattern in the majority of patients. In addition, T2-hyperintense lobules, frequently seen in chondroid lesions, may be seen in 35% of patients. Moreover, skeletal involvement is associated with cortical destruction and extension into the surrounding soft tissue with periosteal reaction in some lesions[11].In the present study, the patient showed similar MRI findings.

Conclusion

The current study shows the first case of extraskeletal mesenchymal chondrosarcoma with metastatic involvement of the iliac vein. ESMC represents a rare entity of highly aggressive tumors with a propensity to metastasize locally and distantly. MRI findings include hypointense lesions on T1W1 and heterogeneously hyperintense lesions on T2WI. Metastases are common and involvement of less commonly reported sites may also occur.

Tables

Table 1. Summary of cases of mesenchymal chondrosarcoma with involvement of the vein

Study	Age(y/o)	gender	Initial site of involvement	Site of metastasis	Radiologic features of the initial lesion	reference
Sabharwal et al.	30 y/o	male	right femoral vein	left pulmonary artery	-	[13]

Zhang et al.	45 y/o	female	Left iliac vein	Local invasion beyond the wall of the vein	Contrast-enhanced computed tomography: a large lobulated mass in the left iliac vein with scattered calcifications	[12]
Simon et al.	25 y/o	female	Left iliac vein	Paravertebral soft tissue	computed tomography scan: heterogeneous lesion with areas of granular, fine calcification T2WI: heterogeneous with fine hypointense foci due to calcification	[20]
Guo et al.	40 y/o	male	Right femoral vein	Pancreas-right upper pulmonary vein- right pulmonary hilum-mediastinal and axillary lymph nodes-lung nodules	Computed tomography scan: Lesion with scattered calcification	[21]
Kim et al.	28 y/o	Female	Left femoral vein	No metastasis or local invasion	Computed tomography scan: multi-lobulated mass with scattered calcification	[22]

Oh et al.	41 y/o	male	pancreas	splenic vein	Computed tomography scan: lobulated, heterogeneously enhancing necrotic mass with numerous areas of coarse calcification	[7]
JUAN HU et al.	61 y/o	female	retroperitoneum	Inferior vena cava	Ultrasound: heterogeneous retroperitoneal masses with increased areas of echogenicity associated with dense posterior shadowing Computed tomography scan: heterogeneous areas with diffuse, dense, ring- and arc-like calcifications T1W1: hypointense lesions T2WI: elevated signal intensity	[9]

Figures description

Figure 1. (A and B) non-contrast-enhanced axial CT images exhibit a heterogeneous retroperitoneal mass with extensive dense popcorn-like calcifications in the left paracervical area and the left common iliac vein. (C and D) Post-contrast images reveal only subtle heterogeneous enhancement of the mass, mainly in its periphery. The lytic bony lesions are identified in left iliac bone, indicative of metastasis. (D) Post-contrast coronal CT clearly displays the extension of the mass to the ipsilateral distended external, internal and common iliac veins.

Fig2. Axial T2 weighted(A) and axial precontrast T1 weighted (B) MR images represent a lobulated heterogeneous left paracervical mass with central popcorn signal voids (calcifications). The coronal STIR (C) and sagittal T2 weighted images can well display the extension of the mass to the expanded ipsilateral internal, external and common iliac veins. Coronal \euro and sagittal (F) contrast-enhanced images revealed peripheral moderate enhancement of the non-calcified components of the mentioned retroperitoneal mass and its

extension to the left iliac vein complex as well as multiple enhancing bony metastases.

Fig 3. Bone scan represents multiple metastatic lesions involving bilateral pelvic bones, both femoral heads, some of the bilateral ribs as well as vertebral bodies.

Conflict of interest

The authors whose names are listed immediately below certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non - financial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter or materials discussed in this manuscript.

Competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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