Malignant Mesenchymoma Presenting as a Paraspinal Mass in a 14-year-old Boy

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ABSTRACT

Malignant mesenchymoma is a rare soft-tissue tumour. We report a case of malignant mesenchymoma in a 14-year-old boy who presented with a rapidly growing mass on his back. Computed tomography revealed a slightly hypodense tumour with heterogeneous contrast enhancement and focal calcification in the right paraspinal region. In T1-weighted magnetic resonance images, the tumour appeared slightly hyperintense relative to skeletal muscle; short TI inversion time inversion-recovery images showed a heterogeneously hyperintense tumour with minimal perilesional oedema. Heterogeneous contrast enhancement was noted in magnetic resonance images after intravenous gadolinium administration. Pathological examination of the resected specimen showed a malignant mesenchymoma with components of leiomyosarcoma and osteosarcoma. This case illustrates that definitive diagnosis of malignant mesenchymoma can be reached only when the tumour is thoroughly sampled and examined.

Key Words: Magnetic resonance imaging; Mesenchymoma/pathology; Soft tissue neoplasms; Spinal diseases; Tomography, X-ray computed

INTRODUCTION

Malignant mesenchymoma is a rare soft-tissue tumour. The disease is generally regarded as a high-grade neoplasm that is characterised by the presence of more than one different mesenchymal tissue component. There is still debate, however, as to the correct terminology of this disease entity. We report a case of malignant mesenchymoma in a 14-year-old boy who presented with a right paraspinal mass to illustrate the imaging features and approaches used in diagnosis of malignant mesenchymoma.

CASE REPORT

A 14-year-old boy presented to the Department of Orthopaedics and Traumatology at the Queen Elizabeth Hospital in June 2003 with a 5-month history of painful swelling in the right paraspinal region of his back after a trivial injury. The size of the swelling had rapidly progressed within these 5 months. There was no constitutional symptom such as weight loss or fever. The patient’s previous health had been unremarkable. Physical examination revealed a firm mass in the right paraspinal region that was fixed to the underlying structure. There was no overlying skin change, no neurological deficit, and no enlarged lymph node or abdominal mass. Haematological and biochemical blood test results were normal.

Non–contrast-enhanced computed tomography (CT) revealed a slightly hypodense 12-cm tumour with focal calcification in the right paraspinal region. After intravenous contrast administration, the tumour showed heterogeneous contrast enhancement and was distinct from the kidney. No bony erosion was visible (Figures 1 and 2). In T1-weighted magnetic resonance imaging (MRI) scans (repetition time [TR], 627 ms; echo time [TE], 15 ms), the tumour appeared as an area in the right erector spinae muscle that was slightly hyperintense mass relative to skeletal muscle (Figure 3). Short TI inversion time inversion-recovery (STIR) sequence images (TR, 7500 ms; TE, 112 ms) showed a heterogeneously
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Hyperintense lesion with minimal perilesional oedema (Figure 4). Heterogeneous contrast enhancement was noted after intravenous administration of gadolinium (Figure 5). Bone scanning with technetium Tc 99m hydroxymethane diphosphonate to stage the tumour showed no increase in vascularity or blood pooling.

Ultrasound-guided biopsy of the paraspinal tumour was subsequently performed at the hyperechoic part of the lesion. Histological examination of the biopsy specimen showed a high-grade sarcoma suggestive of an extraskeletal osteosarcoma. The patient was given neoadjuvant chemotherapy of high-dose methotrexate, cisplatin, and adriamycin. After the first cycle of chemotherapy, however, the mass increased in size. Hence, the patient underwent tumour resection with a wide margin that included tissue from the lower right 3 ribs, spinous processes, transverse processes of T10 to L4 vertebrae, paraspinal muscle, and psoas muscle.

Pathological examination of the resected specimen showed a high-grade sarcoma with various histological patterns in different areas. There was a leiomyosarcoma component that consisted of fascicles of spindle cells displaying mild nuclear atypia, cigar-shaped nuclei, eosinophilic cytoplasm, and occasional mitotic figures (Figure 6a). There was also an osteosarcoma component that consisted of pleomorphic cells, osteoid formation, and calcification (Figure 6b). In most areas, the tumour had a markedly pleomorphic appearance and no apparent lineage of differentiation. Tumour giant cells and frequent mitotic figures were identified in these areas (Figure 6c). Immunohistochemical staining indicated that some of the tumour cells expressed the muscle marker desmin; rarely, cells were also positive for actin. The tumour cells were negative for the skeletal muscle marker myogenin. All these observations suggested a diagnosis of malignant mesenchymoma with

Figure 1. Precontrast axial computed tomogram showing a slightly hypodense tumour with focal calcification in the right paraspinal region that did not involve the right kidney.

Figure 2. Intravenous contrast-enhanced axial computed tomogram showing heterogeneous contrast enhancement.

Figure 3. T1-weighted axial magnetic resonance image (TR, 627 ms; TE, 15 ms) showing a soft-tissue mass, slightly hyperintense to skeletal muscle, in the right erector spinae muscle.

Figure 4. Magnetic resonance image with short T1 inversion time inversion-recovery (TR, 7500 ms; TE, 112 ms) showing a heterogeneously hyperintense tumour with minimal perilesional oedema.

Figure 5. Bone scan with technetium Tc 99m hydroxymethane diphosphonate showing no increase in vascularity or blood pooling.
leiomyosarcoma and osteosarcoma components. Because the tumour had extended to the medial resection margin, the patient received postoperative irradiation therapy. Three weeks after the start of radiotherapy, follow-up MRI showed local tumour recurrence, and CT of the thorax showed multiple secondary lung tumours (Figure 7). The patient refused further chemotherapy and remained asymptomatic at the last follow-up visit, 4 months after surgery.

**DISCUSSION**

Malignant mesenchymoma is a rare malignant soft-tissue neoplasm and was first described by Stout in 1948 as a tumour that is “composed of two or more cellular types, any of which, if taken by itself, might be considered a primary malignant [mesenchymal] neoplasm”. The definition of malignant mesenchymoma is currently controversial. The World Health Organization classification of tumours recommends that the term be applied to “sarcomas that exhibit two or more lineages of specialized differentiation”. Kempson et al suggested that instead of using “mesenchymoma”, one should classify the tumour on the basis of the predominant pattern of differentiation and mention the other pattern. Another approach is to label the tumour as a mixed mesenchymal neoplasm and to specify the lineages of differentiation that are present. Weiss and Goldblum also proposed to abandon the term “malignant mesenchymoma” and to diagnose such sarcomas “by identifying the lines of differentiation, their appropriate amounts, and the grade of the most aggressive component”. Hence, no consensus on the nomenclature has yet been reached.

Malignant mesenchymoma frequently contains three lineages of differentiation: lipoblastic, muscular (smooth or skeletal muscle), and osteocartilaginous. The histogenesis is uncertain and seems to arise from primitive and uncommitted mesenchymal elements that have differentiated along multiple cell lines. In a review by Suzuki et al of 98 malignant mesenchymomas, the most common lines of differentiation were liposarcomatous (64%), chondrosarcomatous (41%), osteosarcomatous (40%), rhabdomyosarcomatous (38%), leiomyosarcomatous (35%), and angiosarcomatous (17%). Among the children studied, the most frequent lines of differentiation were rhabdomyosarcomatous (43%), angiosarcomatous (33%), leiomyosarcomatous (29%), liposarcomatous (29%), chondrosarcomatous (26%), and osteosarcomatous (14%).

Figure 5. Fat-saturated magnetic resonance images after intravenous administration of gadolinium showing heterogeneous contrast enhancement: (a) axial view and (b) sagittal view.

The incidence of malignant mesenchymoma is not well documented in the literature because most of the articles are reports of small case series. In a review of cases of malignant mesenchymoma managed from 1972 to 1994 in the Memorial Sloan-Kettering Cancer Center in the United States, Brady et al identified only 8 cases among 2500 patients with soft-tissue sarcoma. Among adults, males and females were equally affected; however, among children, males were affected more than twice as frequently as females. In the reported cases, malignant mesenchymoma was most commonly found among older patients and occurred infrequently among children and young adults.
The anatomical distribution of malignant mesenchymoma usually involves the retroperitoneum and thigh. Other sites include the upper and lower extremities; anterior and posterior trunk; mediastinum; head and neck region; kidney; small intestine; and peritoneum and mesentery. Furthermore, the orbit, spermatic cord, brain, bone, and retrorectal space can be affected. There are also reports of malignant mesenchymoma in association with congenital lung cyst among children. The disease frequently presents clinically as a fast-growing, large, fixed mass that is palpable in the deep soft tissue; distant metastasis to the lungs occurs within 12 to 18 months. Systemic symptoms can include anorexia, recurrent fever, and night sweats.

Most cases of malignant mesenchymoma are not presented in radiological journals, and there is a lack of detailed description of radiological features of the disease in the literature. Suzuki et al reviewed the radiological features of 5 patients with pathologically proven malignant mesenchymoma and found that in most cases, plain CT or ultrasonography at presentation revealed malignant mesenchymomas that were larger than 10 cm, well circumscribed, and heterogeneous, and that exhibited extensive calcification. Plain CT also showed a fat-dense structure in a tumour in one case.

In general, contrast-enhanced CT showed heterogeneous enhancement, whereas MRI showed essentially hypodense areas on T1-weighted images and heterogeneous hyperintense areas on T2-weighted images. Hence, Suzuki et al suggested that a large tumour size, a sharp margin, heterogeneous composition, and massive calcification are all typical findings of malignant mesenchymoma. Depending on its content, malignant mesenchymoma may demonstrate various radiological features. If a large retroperitoneal soft-tissue mass is present, together with several different sarcomatous features (e.g., prominent calcification or ossification, or a fatty component), the differential diagnosis should include malignant mesenchymoma.

The radiological differential diagnosis includes primary extra-osseous osteosarcoma, metastatic osteosarcoma, myositis ossificans, teratoma, and calcified haematoma.

Figure 6. Photomicrographs showing different lineages of differentiation. (a) Leiomyosarcoma component: fascicles of spindle cells displaying mild nuclear atypia, cigar-shaped nuclei, eosinophilic cytoplasm, and occasional mitotic figures (arrow); (b) osteosarcoma component: pleomorphic tumour cells with osteoid deposition (arrows); and (c) area with no apparent line of differentiation: markedly pleomorphic tumour cells including tumour giant cells (arrowheads) with frequent mitotic figures (arrow) [haematoxylin and eosin; original magnification, x60].

Figure 7. Postoperative follow-up computed tomogram of the thorax in the lung window setting showing multiple secondary lung tumours.
In most of the reported cases, the diagnosis of malignant mesenchymoma was made from the surgically resected specimen. There are also reports of preoperative needle biopsy that show only one element of the sarcomatous component. Surgical removal — especially complete resection — is regarded as the preferred treatment. The effect of chemotherapy or radiation has not been evaluated in a large study, but in general, these interventions are not considered to be efficacious. Although adjuvant radiation therapy may reduce the incidence of local recurrence, it may not influence the overall survival.

The prognosis of malignant mesenchymoma is controversial according to studies in the early literature. Nevertheless, this disease entity is currently accepted as a high-grade malignant neoplasm. Newman and Fletcher suggested that it displays low-grade malignant behaviour, but this conclusion was based on only 6 cases in which the median follow-up duration was 4.2 years. The prognosis also depends on the prevailing mesenchymal component; the prognosis is most favourable when liposarcomatous components prevail, and very poor when rhabdomyosarcomatous components prevail. Localised nodal metastasis is just as ominous as distant pulmonary metastasis. And although a rapidly growing tumour has a pronounced tendency to recur, such a tumour only occasionally metastasises.

In our case, it was prudent to consider malignant mesenchymoma as one of the possible diagnoses after biopsy confirmed a soft-tissue sarcoma with intraloskeletal radiological features that showed different tissue characteristics. Sampling from multiple sites with different radiological features may be required to establish a preoperative diagnosis of malignant mesenchymoma, but this approach may not be practical in some instances. It is very understandable that a definitive diagnosis of malignant mesenchymoma can be reached only when the tumour is thoroughly sampled and examined. However, because the prognosis is frequently determined by the prevailing component, the treatment planned after fully studying the excised specimen may not be very different from that planned after the initial biopsy.

REFERENCES


