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CASE REPORT

Malignancy is a rare but serious cause of low back pain especially in young people

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ABSTRACT

This is true that many cases of acute low back pain are caused by radicular pain due to discopathy, however, the history and physical examination in low risk patients as well as continuous radiography usually provide clues to the rare but potentially serious causes of low back pain such as malignancy. The patient is an 18-year-old man who referred to an orthopedist for the first time three months prior to the study with a complaint of a backache and received out-patient treatment. One month ago he visited a doctor for lack of improvement and aggravation of the symptoms. In X ray imaging from pelvis performed three months earlier, nothing was found except for a lesion that was probably differentiated from intestinal gas and not reported. However, by repeated X-ray a month later and because of lack of relocation of lesion created high doubt ex parte lytic lesion for the patient, A biopsy specimen was taken from this site. The primary result indicated sarcoma, while after immunohistochemistery, final result revealed peripheral malignancy nerve sheath tumor for patient. Malignant peripheral nerve sheath tumor (MPNST) are rare malignancies from the origin of peripheral nerve sheath cells. These tumors almost present as an enlarging mass from a peripheral nerve root. We should consider the importance of low back pain in a patient that does not have serious risk factors and inflammatory or mechanical nature of the pain because in some cases low back pain can indicate severe diseases.

Key words: neurofibromatosis type one, discopathy, radicular low back pain, malignant peripheral nerve sheath tumor (MPNST)

INTRODUCTION

Acute low back pain is a common cause for referring to doctors. Although there is a large differential diagnosis, the definite etiology is rarely diagnosed, despite musculoskeletal events are usually suspected. In many patients, there is clue to radicular symptoms or a basic systemic disease.

The aim of this study is to evaluate the importance of low back pain in young people especially those with no risk factor. This is true that many cases of acute low back pain are caused by radicular pain due to discopathy, however, the history and physical examination in low risk patients as well as continuous radiography usually provide clues to the rare but potentially serious causes of low back pain such as malignancy.

CASE REPORT

The patient is an 18-year-old man who referred to an orthopedist for the first time three months prior to the study with a complaint of backache and received out-patient treatment.

One month ago he visited a doctor for lack of improvement and aggravation of the symptoms.

The pain radiated to left and outer part of the calf and the heal region. This pain was accompanied by claudication and not depended on exertion. It deteriorated by rest especially in the morning. His vital signs were recorded as blood pressure of 125 on 65 mmHg, pulse rate of 73 beats per minute, respratory rate of 17 beats per minute; and tempreture of 37.3 ºC.

No pathologic sign was observed in the abdomen, heart, and lung.

According to extremities examination, right and left

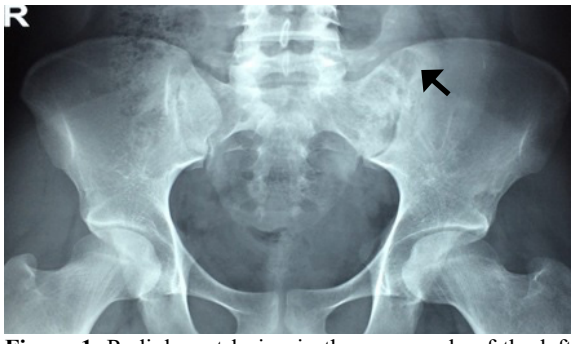

Figure 1, Radiolucent lesion in the upper pole of the left iliac bone.

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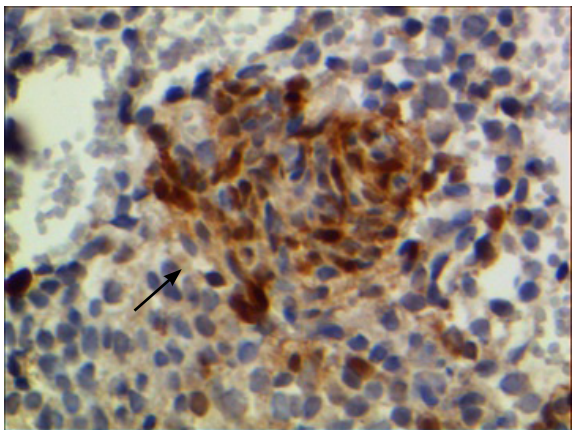

Figure 2, Immunohistochemical staining for S-100 protein was positive in tumor cells (arrow).

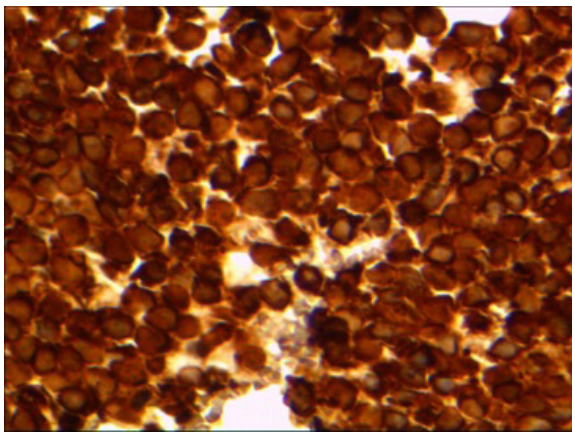

Figure 3, Positive vimentin staining in the spindle -shaped cells.

superior, and right inferior extremities were normal.
In left inferior extremities examination abnormal gait was observed.
In left inferior extremity in 30 degrees, The straight leg raise (SLR) test and Patrick (Faber) test were both positive.
(Ex parte involvement of left sacroiliac joint)
In comparison with right inferior extremity, Deep tendon reflex (DTR) decreased in left inferior extremity and its muscular force was approximately 4/5.
In X ray imaging from pelvis performed three months earlier, nothing was found except for a lesion that was probably differentiated from intestinal gas and not reported. However, by repeated X ray a month later and because of lack of relocation of lesion created high doubt ex parte lytic lesion for patient (Fig. 1).
Spiral computed tomography was requested from lumbar and hip section which showed destruction of mass lytic and wide transitional zone with soft tissue component expanded to left neural foramen (70 × 55 mm). A biopsy specimen was taken from this site.
Primary result indicated sarcoma, while after immunohistochemistry (IHC), final result revealed peripheral malignancy nerve sheath tumor for patient requested ancology cosultion that don't found any special lesion Ex parte metastasis in abdominal, thoracic and pelvic computed tomography.

DISCUSSION

MPNSTs are rare malignancies from the origin of peripheral nerve sheath cells. These tumors almost present as an enlarging mass from a peripheral nerve root of the trunk (~50%) in the extremities (~30%) or the head and neck region (~20%).2 Infiltration in side bone in a paraspinal MPNST is a known process, although a primary intraos¬seous MPNST is rare. An intraosseous MPNST may grow from minute, mainly unmyelinated nerve roots that coexist with nutrient vessels and branch in Volkmann's canals and bone marrow.[5,6] To the best of our knowledge, only 19 cases of intraosseous MPNSTs, including the present study, have been reported in the literature to date.[3,4,7,8] There were 9 male and 10 female patients, and the age at diagnosis varied from 4 to 76 years. A total of 8 out of 19 cases (42%) had the disease in the mandible or maxilla while 6 (32%) had the disease in a vertebral body of the spine.
Despite <50% of MPNSTs arise in patients with neurofibromatosis type one (NF1)[8], intraosseous MPNST cases were not connected with the hereditary NF1 syndrome, except for one case. The case of the present study was not connected with NF1 either. Due to a wide morphologic feature and lack of certain markers, the diagnosis of intraosseous MPNST was very difficult, especially in a case lacking the signs of NF1 and classic histopathological markers. High grade MPNSTs may be similar to other malignancies, including synovial sarcoma, fibrosarcoma, and malignant fibrous histiocytoma. In the present study, the histological diagnosis of the lumbar mass was MPNST with positive IHC markers for smooth muscle actin, vimentin, S 100 (Figs. 2,3) and Ki67, while negative for EMA, AE1/AE3,CD99 ,CK7, CD20, CD45, pancytokeratin, Nse, Chromogranin A and Desmin.
Several antigens are important in identification of nerve sheath differentiation, including myelin basic protein, S 100 protein, and Leu 7. The protein S 100 is the most commonly used antigen for neural distinction and can be recognized in ~50% of MPNSTs, although the staining is typically local and limited to a several number of cells. By experience, an MPNST with diffuse immunoreactivity for the S 100 protein is unusual, such a staining pattern always shows that other benign diagnoses should be reviewed, especially cellular schwannoma. Leu 7 and myelin basic protein have been identified in ~50 and ~40% of MPNSTs, respectively.[9]
The molecular pathogenesis of MPNST remains unclear. In contrast to several other sarcomas, there was no responsible chromosomal translocation and standard karyotypes without specific aberrations.[1] However, losing chromosome 17q, the location of the NF1, has been recognized in 25 50% of reported sporadic and NF1 related cases, usually in the form of chromosomal monosomy. As the same, NF1 remotion

Table 1, Summary of reported cases of intraosseous MPNST.

Case (ref.)	Age (years)	Sex	Location	NF1	Surgery	Adjuvant therapy	Outcome (months)
1 (17)	55	M	Ulna	-	CR	None	NED (20)
2 (18)	65	F	Mandible	-	NA	NA	NA
3 (19)	65	M	Mandible	-	Resection	RT	AWD (NA): recurrence
4 (20)	4	F	Mandible	NA	NA	NA	NA
5 (21)	28	M	Ulna	-	NA	NA	NA
6 (22)	11	F	Mandible	-	Resection	None	NED (6)
7 (23)	76	F	Mandible	-	NA	NA	NA
8 (24)	61	F	Maxilla	-	NA	NA	NA
9 (25)	47	F	Maxilla	-	Resection	RT CT	DOD (22)
10 (26)	50	M	Mandible	-	NA	RT	DOD (12)
11 (27)	28	M	Femur	-	Resection	None	DOD (1): pulmonary metastasis
12 (28)	26	M	Femur	-	Resection	CT	DOD (15): pulmonary metastasis
13 (29)	29	M	Ulna	-	Resection	None	DOD (36): pulmonary metastasis
14 (6)	40	F	C2	NA	STR	PSF	DOD (12): pulmonary metastasis
15 (7)	59	F	T3	-	STR	PSF RT CT	AWD (46): bone metastasis
16 (8)	75	F	T7	-	STR	PSF RT	DOD (6): pulmonary metastasis
17 (9)	41	M	C7	-	CR	PSF CT	NED (24)
18 (10)	75	M	L3	-	STR	NA	NA

MPNST, malignant peripheral nerve sheath tumor; NF1, neurofibromatosis type 1; M, male; F, female; C, cervical; T, thoracic; L, lumbar; CR, complete resection; NA, not available; STR, subtotal resection; PSF, posterior spinal fusion; RT, radiotherapy; CT, chemotherapy; NED, no evidence of disease; AWD, alive with disease; DOD, dead of disease.

in Fluorescence in situ hybrization (FISH) analysis may help in differentiating MPNSTs from other high grade malignancies with coinciding morphological characters. Perry *et al*.[10] showed that NF1 remotions were detectable in 76% of sporadic and NF1 related MPNSTs by FISH analysis and detected in five out of six high grade MPNSTs that were without S 100 protein immunoreactivity, being a significant marker for Schwann cells. furthermore, it has been reported that of eight cases with MPNST, NF1 remotion was discovered within the S 100 positive cellular accumulations of four MPNSTs (50%), while S 100 negative core were observed in all eight MPNSTs. These results indicated the prevalence of NF1 remotion in MPNSTs, indifferent of S 100 protein expression.[11] Complete surgical resection with negative margins is considered as the best treatment for MPNSTs, in a study by Wong *et al*.[12]

A hundred and twenty eight patients with MPNSTs were observed and 83% of them underwent en bloc resection, of whom only 48% had negative surgical margins. Due to the surrounding spinal cord, dura mater and large blood vessels many cases became compli¬cated in en bloc resection of spinal MPNST. Adjuvant therapy like radiation to a dose of >60 Gy can help to control local disease.[13] The usefulness of chemotherapy for MPNST (with doxorubicin use alone or in combination with other drugs[14] remains controversial.

CONCLUSION

The present study introduces a case of intraosseous MPNST arising in a lumbar vertebra that the patient is candidate for *en bloc* resection of the tumor and adjuvant chemotherapy. Since the outcome remains poor, further studies are required on genetic therapy of the tumor based on its molecular pathogenesis. The other point that we should consider is the importance of low back pain in a patient that does not have serious risk factors and inflammatory or mechanical nature of the pain, because in some cases low back pain can indicate severe diseases.

CONFLICT OF INTEREST

None.

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