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Desmoplastic fibroma of the cervical spine

Received: 12 September 2004
Revised: 11 November 2004
Accepted: 29 December 2004
Published online: 4 June 2005
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Abstract There have been only a few cases of desmoplastic fibroma of the spine in the literature and only one of them was purely located on the cervical spine. We report a new patient with the diagnosis of desmoplastic fibroma of the fourth cervical spine. The patient had the complaints of left arm and neck pain. After his radiological evaluation, a mass lesion was found on the left lamina of the fourth cervical spine. Surgical treatment was performed, and the histopathological examination revealed the diagnosis of desmoplastic fibroma. Patients with desmoplastic fibroma of the cervical

spine may present with the arm and neck pain mimicking cervical disc disease. Higher index of suspicion by the clinicians must be practiced to make the appropriate diagnosis. Successful surgical outcome may be achieved in these patients.

Keywords Cervical spine · Desmoplastic fibroma · Surgery

Introduction

Cervical spine is an unusual location for desmoplastic fibroma which is a benign but locally aggressive tumor of the connective tissue. It's very uncommon and usually arises in the mandible, femur or humerus [2].

A 41-year-old man with desmoplastic fibroma of the fourth cervical spine was treated by complete resection. There was no recurrence in the following controls after the surgery. To our knowledge, this appears to be the second case report of such a diagnosis; and therefore, probably represents a rare and unusual clinical situation. We reported the clinical, radiological and surgical features of this rare phenomenon and reviewed with the literature.

Case report

A 41-year-old man was admitted to our department with the complaints of headache, and neck pain radiating to

the left arm with numbness for 2 months. The pain was slowly increasing and was unresponsive to analgesics. On physical examination, local tenderness was observed in the C5 area. There was no changes in motor functions. Laboratory examinations did not reveal any abnormal finding. The plain roentgenography of the cervical spine showed minimal thickness on the left lamina of the fourth cervical vertebra. The computed tomography (CT) of the cervical spine was done and mass lesion causing enlargement of the left lamina and spinous process of the fourth cervical vertebra was detected (Fig. 1). After this finding, we performed cervical spine magnetic resonance imaging (MRI) with a 1.5 T unit, using spin-echo technique with T1 and T2 and post-Gadolinium T1-weighted sequences. The aim of the MRI study is to evaluate any compression of the spinal cord and to determine the soft-tissue expansion if it exists. On the MRI, both of the spinal cord and the cervical roots were intact and no soft-tissue mass lesion was detected. The possible cause of radiating pain to the

arm may be a minor nerve root irritation which was not visible in CT or MRI scans. The differential diagnosis were non-ossifying fibroma, giant cell tumor and fibrous dysplasia before treating the patient. Biopsy was not performed before the definitive surgery because of the risk of injury to nearby neural tissue.

The patient underwent surgery with the left laminectomy on the fourth cervical vertebra. During operation, thickened left lamina of the fourth cervical vertebra was seen and we achieved complete resection of the mass lesion expanding the lamina. The spinous process was also excised due to risk of microscopic invasion of tumor. The contralateral lamina was preserved.

The pathological examination of the specimen showed desmoplastic fibroma of the connective tissue. The lesion is composed primarily of dense collagenous tissue, and the fibroblasts form obtuse bands without mitotic activity and fibrosis. These fibroblasts are embedded in the dense collagenous matrix. Absence of nuclear anaplasia confirmed the diagnosis of desmoplastic fibroma (Fig. 2).

The patient was followed at our clinic in the post-operative period and discharged without any complication. The follow-up cervical CT scan was consistent with the lack of recurrence at the end of 4 years after surgery (Fig. 3).

Fig. 1 Preoperative axial CT scan of the patient showing tumor located in the lamina of the fourth cervical vertebra. The thin cortex was observed

Discussion

Desmoplastic fibroma is a benign but locally aggressive intraosseous neoplasm that was first described in 1958 by Jaffe [5]. It's a tumor of fibrous connective tissue origin marked by the formation of collagen. Sparse cellularity, lack of pleomorphism, and decreased mitotic activity characterize this tumor.

Desmoplastic fibroma is a very rare tumor which arises usually in the mandible, femur, humerus or pelvis; but rarely involves the calvaria or spine. The spinal location is extremely rare and only a few cases have been previously reported in the literature. Most of them were located in the thoracic or lumbar level [4, 6, 7, 9–11]. Our patient is the second case in the literature with desmoplastic fibroma of the cervical spine. The first was the desmoplastic fibroma of the axis which was reported by Fuji et al. in 1988 [3]. That was a 24-year-old-male patient who had complained of clumsiness of the hand and gait disturbance. The posterior part of the axis was totally destroyed by tumor in this case. They performed complete resection with posterior spinal fusion for this benign spinal tumor. Aboulafia et al reported a case of desmoplastic fibroma located in the cervico-thoracic junction (C7-T1). They did not perform surgery and the diagnosis was confirmed with CT-guided needle biopsy [1]. In our opinion, the biopsy may pose a risk of nearby neural tissue damage and complete resection is required for diagnosis and treatment.

The differential diagnosis of the desmoplastic fibroma is a matter of debate today. Therefore, it's too difficult to distinguish from the other bony lesions such as giant cell tumor, chordoma and fibrous dysplasia of the spine. Histologically, the primary low-grade fibrosarcoma of bone is the most important and difficult tumor in the differential diagnosis [3]. It can also involve pelvis and calvaria, but it's extremely rare.

Computed tomography of the spine is the procedure of choice in the bone tumors of the spine. Radiologically, these tumors have well-defined lytic and expansile lesions with a soap-bubble appearance. Thinning of the overlying bony cortex in the absence of a periosteal reaction is common. Sclerotic reaction at the margins is mild to absent. Our case shows the characteristic radiological features of desmoplastic fibroma; well-defined, expansile, osteolytic lesion but without soft-tissue mass and spinal cord compression. The left lamina of the fourth cervical spine was enlarged with the hypodense mass lesion and the margins of the lamina became very thin. The magnetic resonance imaging of this tumor is useful in determining the presence of spinal cord compression. Kong et al. [6] reported a case of lumbar spinal desmoplastic fibroma, and mentioned the MRI features of this rare tumor. In their case, a 24-year-old male patient presented with low back pain radiating to both

Fig. 2 The histopathological appearance of the tumor. The tumor was composed of interlacing fascicles of the low fibroblastic cellularity. Mitotic activity is absent (Hematoxylin and eosin, $\times 100$)

the lower extremities. The tumor was located at the vertebral body of L5 level and he underwent total corpectomy with posterior fusion.

Desmoplastic fibroma is a locally aggressive tumor with a 20–30% recurrence rate reported; therefore, complete excision is recommended. Local excision and curettage with cryotherapy may be adequate for limited

lesions and those inaccessible to complete resection. Radiation therapy is of no benefit in these tumors [8].

To conclude, desmoplastic fibroma is a benign tumor of the connective tissue and may seldom present with neck pain. These tumors are easy resectable if the complete excision is performed. Otherwise, local recurrence may occur, and the treatment may become difficult.

Fig. 3 Postoperative axial CT scan showing the absence of tumor

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