Superior mediastinal chordoma presenting as a bilobed paravertebral mass

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Abstract

Thoracic chordomas are rare, low-grade malignant tumours arising from the notochordal remnants. These tumours are locally invasive and hence have a tendency for frequent local recurrence. This article presents an unusual appearance of a chordoma, as a bilobed tumour involving both the hemithoraxes. Since the treatment of choice for these tumours is complete surgical excision this involved multiple surgical procedures.

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1. Introduction

Chordomas are slowly growing malignant tumours arising from the remnants of the notochord. Normally the notochord persists only in the nucleus pulposus of the intervertebral disc. Chordoma is the only embryonic neoplasm that presents in the later decades of life, typically in the fourth to fifth decades.

The most prevalent site for chordomas are the sacrococcygeal region (50–55%), craniooccipital region (25–30%), cervical spine (8%) and lumbar spine (5%). Thoracic chordomas account for only 1–2% of all chordomas [1]. However, chordomas presenting as a soft tissue mass in the posterior mediastinum without involvement of the vertebral bodies are rare [2] and a bilobed chordoma involving both the hemithoraxes has not been described so far.

2. Case report

A 33-year-old woman, while being investigated for a routine gynaecological procedure, was found to have on chest X-ray a bilobed paravertebral mass in the upper mediastinum. Although the mass was an incidental finding, she subsequently developed swelling and redness of the face on lifting her hands to dress her hair and also a tingling sensation along the medial side of the left forearm. Physical examination did not reveal any abnormality.

2.1. Investigations

Chest X-ray (Fig. 1) showed a well-defined homogeneous bilobed mass, each half measuring about 5 × 5 cm in the upper mediastinum. Computed tomography (Fig. 2) revealed a fairly symmetrical bilobed, lower attenuation, non-enhancing mass in paraspinal region of the superior mediastinum. There were a few small areas of calcification. The spine was normal.

2.2. Operation

She underwent an excision biopsy through a right posterolateral thoracotomy. The lung was normal and no lymphadenopathy was noted. The mass was 4 × 5 cm in size and entirely extrapleural being adherent to the second and third thoracic vertebrae. This mass had a myxoid/gelatinous appearance and was enclosed in a capsule of varying thickness. It was relatively avascular and there was no obvious feeding vessel. This pole of the mass was excised almost completely. The isthmus remained adherent to the anterior spinal ligament, the vertebral body and fibres of the erector spinae muscle. Postoperative computed tomography showed that there was a 1 × 2 cm residual mass anterior to the spine and a 4 × 5 cm mass on the left side. The patient had an uneventful postoperative period.
2.3. Histopathology

The tumour was a chordoma. The tumour was partly encapsulated and divided into nodules by fibrous septae, contained abundant mucoid matrix separating large tumour cells with clear cytoplasm (physaliferous cells). There was mild nuclear pleomorphism and the mitotic rate was low. Immunohistochemical stains showed strong cytoplasmic reactivity to cytokeratin MNF116, epithelial membrane antigen and S-100 protein.

2.4. Re-operation

The patient was subsequently re-admitted for a second operation. She underwent a left posterolateral thoracotomy. The thoracic duct and oesophagus were separated from the anterior surface of the tumour. The mass was excised along with the adjacent periosteum and the muscle fibres of the erector spinae. Although a good clearance was obtained a post resection computed tomography (CT) scan confirmed that a portion of the isthmus of the tumour was still present on the right side. Subsequently she was seen by the oncologists and it was felt that radiotherapy presented a significant risk to adjacent vital structures. A third procedure was undertaken to excise the remnant of tissue together with the adjacent fibres of erector spinae and anterior spinal ligament. A good clearance was achieved.

The patient had an uneventful postoperative period. She had no further swelling or redness of her face on lifting her hands which was thought to be due to venous compression by tumour in the thoracic inlet (Pemberton’s sign). CT scan performed 6 months after the last surgery has not shown any recurrence.

3. Discussion

The first chordoma-like lesion was described by Lushka in 1856 as a small, soft, jelly-like tumour. A year later Virchow described the tumours as degenerated cartilage and called them as ecchondrosis physaliphora. During the same period Muller suggested that the tumours might be of notochordal origin. In 1895 Ribbert was able to experimentally establish that the tumours developed from notochordal remnants and he coined the word chordoma [3]. In 1951 Crowe and Muldoon reported the first case to undergo surgical excision of a thoracic chordoma [4]. Despite current evidence indicating that chordomas contain immunophenotypic and ultrastructural markers of epithelial differentiation, they are usually classed among tumours of bone, because of the extensive involvement and destruction of adjacent bony structures [2].

As no large series of thoracic chordomas have been reported, incidence and survival figures are uncertain. For all chordomas there is a 2:1 male to female ratio and the highest incidence is found in the fourth to fifth decades. The differential diagnosis of mediastinal chordomas include neurogenic tumours or cysts (neurofibroma, schwannoma, ganglioneuroma, neuroblastoma and neuroenteric cysts), and less commonly aneurysms of the arch and descending aorta, diaphragmatic hernia, lymphadenopathy and extra-medullary haematopoiesis. Preoperative CT and magnetic resonance imaging are useful in evaluating the extent of the tumour. Fine-needle biopsy is often not contributive [5].

Chordomas are considered as low-grade malignant lesions because of their tendency for local invasion and frequent local recurrence with destruction of adjacent vertebrae and nerve trunks. They metastasize late in their course. However, the presence of metastasis often does not alter the prognosis of patients since death in most cases is from local recurrence.

The treatment of choice for chordomas is en bloc surgical excision with adequate surgical margins. Initial reports had implied that adjuvant radiotherapy was ineffective. Recent
reports with higher dose radiotherapy have more encouraging results. Tumour shrinkage or resolution has also been demonstrated with the use of interstitial implants. Adjuvant radiotherapy should be given to all patients with postoperative residual tumours [6]. The prognosis for these tumours is unpredictable and depends on the location and extent of resection.

The initial procedure was undertaken to establish a histological diagnosis and to excise the mass. Because of involvement of the anterior spinal structures, the clinical impression was that further treatment would not be surgical and an excision biopsy of the right-sided mass was performed. When the diagnosis of chordoma was established, complete excision required two further thoracotomies and limited follow-up has not shown any recurrent disease. With hindsight it might have been possible to limit to two thoracotomies (possibly at the same sitting, but without the benefit of histology).

**References**