A Parapharyngeal Soft Tissue Chordoma Presenting with Synchronous Cervical Lymph Node Metastasis: An Unusual Presentation

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Abstract A 63 year old male presented with a three month history of dysphagia, neck swelling and an oropharyngeal swelling on examination. Initial fine needle aspiration cytology and magnetic resonance imaging (done at a peripheral hospital) suggested a salivary gland neoplasm with lymph node metastasis. An infra-temporal approach was employed to excise the tumour mass and a modified radical neck dissection undertaken to remove the cervical metastasis. Histopathological examination with immunohistochemistry confirmed this to be a soft tissue chordoma. To the best of our knowledge, this is the first documented report of an extra-axial soft tissue chordoma presenting with synchronous metastatic disease. Though rare, this adds to the list of differential diagnoses for complex parapharyngeal lesions. A multidisciplinary approach between head and neck surgery, histopathology, radiology and sarcoma teams is paramount for arriving at the correct diagnosis and to deliver optimal treatment.

Keywords Chordoma · Soft tissue · Head and neck · Parapharyngeal · Metastasis

Introduction

Chordomas are slow growing locally aggressive malignant tumours arising from the embryonic remnants of the notochord along the neuroaxis (e.g. clival/skull base and sacrococcygeal) and account for 1 % of all primary malignant bone tumours [1, 2]. Extra axial chordomas are rare, usually arising in long bones of extremities and are morphologically similar to their axial counterpart [2]. Chordomas rarely arise within soft tissue and are extremely uncommon in head and neck surgical practice [3]. Rarely, skull base chordomas may occur in an extraosseous location and present as a nasopharyngeal mass with an associated clival sinus tract [4, 5]. Only a few cases of soft tissue chordomas have been reported in the head and neck region involving the lateral nasal wall [6], paranasal sinuses [7, 8], oropharynx [9], parapharyngeal space [10], and soft tissues of the neck [11–13]. We present a case of a primary soft tissue chordoma involving the parapharyngeal space with cervical lymph node metastasis. To our knowledge, an extra-axial soft tissue chordoma with synchronous regional metastasis at presentation has not been described in the literature previously.

Case History

A 63 year old male presented to the local Ear, Nose and Throat (ENT) clinic with a three month history of a progressively enlarging right sided neck lump, dysphagia and intermittent right otalgia. A large, smooth and non-tender swelling was seen in the right tonsillar region without mucosal ulceration. In addition, a firm, mobile mass (5 × 5 cm) was also evident in the right cervical level-II region. Initial Fine Needle Aspirate Cytology (FNAC) of

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Fig. 1 MRI (coronal view) shows a discrete mass in the parapharyngeal space extending into the skull base and oral cavity

Fig. 2 Intra-operative photograph showing removal of the tumour from the parapharyngeal space

the neck node suggested the possibility of a metastatic salivary gland neoplasm. Magnetic resonance imaging (MRI) showed a well-defined and large parapharyngeal mass eroding the prevertebral fascia over the second cervical vertebra. This mass appeared separate from the deep lobe of parotid gland (Fig. 1).

Following discussion at the regional head and neck multidisciplinary team (MDT) meeting, the lesion was surgically removed using a transcervical–transpharyngeal, infra-temporal approach with a mandible split (Fig. 2). Intraoperatively, the tumour appeared well encapsulated

and centered in soft tissue with a smooth surface and extending to the skull base showing minor erosion of the anterior surface of the upper cervical vertebra, but without intracranial extension. The patient had a right modified radical neck dissection for removal of metastatic tumour at the time of removal of the main mass.

Histological examination of the main tumour mass revealed cords and lobules of vacuolated physaliphorous cells showing moderate nuclear atypia in a muco-myxoid background (Fig. 3a, b). Immunohistochemistry showed expression of cytokeratins Cam5.2 (Fig. 3c), CK19 (Fig. 3d), MNF116, AE1/AE3, and CK5/6 (not shown). Focal S100 staining was seen in univacuolated tumour cells and scattered adipocytes (Fig. 3e). There was no expression of EMA and no evidence of cartilage or ductal differentiation. The differential diagnosis included a parachordoma or a soft tissue chordoma. The metastatic neck level II deposit showed similar histological features confirming nodal metastasis of the tumour (Fig. 3f). Brachyury staining was performed which showed strong nuclear staining in the primary (Fig. 4a) and metastatic tumour (Fig. 4b) confirming the diagnosis of an extra-axial soft tissue chordoma.

As the primary tumour had intralesional margins, adjuvant post-operative radiotherapy (60 Gray, 30 fractions) was given and the patient was followed up at the ENT clinic at the referral hospital. Four years after the initial surgical and oncological intervention, the patient developed sudden onset neck pain which did not respond to analgesia. A cervical spine X-ray showed C1/2 abnormality and a soft tissue mass. MRI and CT scans showed a pathological fracture of the odontoid process with anterior subluxation and narrowing of the spinal canal secondary to a large soft tissue mass surrounding the body and lateral mass of C2. No evidence of lymphadenopathy or metastatic infiltration was seen elsewhere. An Aspen collar was placed and following an urgent discussion at the sarcoma MDT, a palliative approach was undertaken involving occipital stabilization plates and further radiotherapy.

Discussion

Extra-axial chordomas were first described by Laskowski as ‘*chordoma periphericum*’ in 1951 with a postulated origin from specialized synovial cells [13]. Dabska coined the name ‘*Parachordoma*’ and described the morphological similarity to chordoma [7]. According to the current World Health Organization (WHO) tumour classification of soft tissue and bone, parachordomas are grouped with mixed tumours and myoepitheliomas of soft tissue, and are thought to be related to mixed salivary gland tumours [2]. However, parachordomas are a separate entity from soft

Fig. 3 Photomicrographs showing physalipherous cells within a myxoid stroma as seen with H&E stain (**a, b**); original magnification $\times 100$. Immunohistochemistry showing strong staining for Cam5.2 (**c**) and CK19 (**d**) whereas S100 (**e**) was expressed by adipocytes with focal staining in univacuolated tumour cells; original magnification

$\times 200$. Metastatic lymph node deposit showing similar histological features to the primary tumour (**f**); original magnification $\times 100$. Staining was also seen for MNF116, AE1/AE3, and CK5/6 (results not shown)

tissue chordomas due to their characteristic immunohistochemical and cytogenetic profiles [1]. It is important to differentiate soft tissue chordomas from its histologic mimics such as parachordoma, soft tissue myoepithelioma, and myxoid chondrosarcoma due to the difference in treatment and prognosis.

Recently, it has been proposed that tumours occurring in bone and soft tissue which are morphologically similar to axial chordomas should be classified as extra-axial chordomas if they are immunoreactive for Brachyury and CK19 [1, 14–16]. Brachyury encodes a transcription factor involved in formation of posterior mesoderm and

axial development. It has also been suggested that tumours not expressing these antigens should be categorized as mixed tumours/myoepithelioma/parachordomas [1]. Axial chordomas are reported to have a metastatic incidence range between 5 and 40 % with metastases thought to arise after local recurrence. The current accepted treatment approach for managing chordomas involves wide local excision of the tumour followed by high dose radiotherapy. The main aim for this treatment modality is palliation with curative intent due to high recurrence rates and a poor 5–10 years survival rate [15–17]. However, it is not clear whether this is related to the

Fig. 4 Photomicrographs showing immunohistochemical expression of Brachyury. Primary tumour (a); original magnification $\times 100$, metastatic tumour (b); original magnification $\times 200$

size of the initial lesion or the difficulty in appropriate removal due to complex anatomy of the involved regions. There have been previously reported metastases of soft tissue chordomas during follow-up [18, 19]. However, these were reported before the recently recommended diagnostic guidelines and without Brachyury staining. Lauer et al. [16] recently conducted a clinicopathologic analysis of 11 soft tissue chordoma cases, three of which developed lung metastases during follow-up. To our knowledge, synchronous presence of nodal metastasis at presentation for extra-axial lesions has not been previously reported.

Recent studies have shown superior local control rate using carbon ion re-irradiation and proton therapy in recurrent skull base chordomas [20, 21]. Studies have demonstrated that activated epidermal growth factor receptor (EGFR) along with the signal transducers and activators of transcription (STAT3) pathway are constitutively activated in chordomas and their level of expression may serve as a predictor for prognosis [22, 23]. Downregulation of micro-RNAs 34a and 608 has also been shown to regulate EGFR and the receptor tyrosine kinase (MET) in chordomas. A correlation between Brachyury expression and chromosomal copy aberration (2p) has also been reported [24, 25]. Blockade of these micro-RNAs, STAT3 pathway, and Brachyury may therefore represent potential future molecular therapeutic targets.

The complex anatomy of the head and neck region makes wide excision of malignancies complicated. We recommend clinical and radiological examination for lymph nodes in all cases of soft tissue chordomas to rule out metastasis which can potentially influence the initial surgical plan. Furthermore, due to rarity of these lesions and the complex diagnosis and follow up, it is recommended that such cases are followed up routinely at a specialist sarcoma center.

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