Sarcoma Research - International

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Case Report

Huge Parapharyngeal Extra-Skeletal Mesenchymal Chondrosarcoma with Intracranial Extension: An Exceptional Clinical Entity

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Received: February 29, 2016; **Accepted:** March 24, 2016; **Published:** March 28, 2016

Abstract

Parapharyngeal extra-skeletal mesenchymal chondrosarcoma is an extremely rare highly malignant tumor with a high incidence of local recurrence as well as distant metastasis. In addition, its intricate anatomical location results in subtle clinical features and hence the diagnosis is delayed. Besides, the surgical approach to such a location for complete tumor resection becomes challenging for the surgeon. We report an exceptionally rare case of extra-skeletal mesenchymal chondrosarcoma of parapharyngeal space and discuss the approach to such tumors to add to the current knowledge of this disease.

Keywords: Parapharyngeal; Chondrosarcoma; Malignant; Trans cervical; Extra-skeletal

Introduction

Primary parapharyngeal space tumors are rare representing only 0.5% of head and neck neoplasms, chondrosarcoma is even rarer [1,2]. Parapharyngeal space mesenchymal chondrosarcoma is a highly aggressive malignant tumor having a peculiarity of locoregional or distant metastasis. In addition, its intricate anatomical location results in subtle clinical features and hence the diagnosis is delayed. Besides, the surgical approach to such a location for complete tumor resection becomes challenging for the surgeon. Moreover, a very few cases of parapharyngeal malignancy are available in English literature to provide a clear guideline of management and to asses prognosis.

Therefore, we report here an exceptionally rare case of extra-skeletal mesenchymal chondrosarcoma of parapharyngeal space with intracranial extension in young male and its management.

Case Presentation

Patient history

A 30 year old male presented with progressive lump in right side of neck since 1 year and diplopia since 1 month. He had history of surgery for these 6 months back with excision of right submandibular mass, histopathological examination of which revealed pleomorphic adenoma.

Local examination

There was a diffuse, firm, non tender and fixed swelling of about 8 x 2cm posterior to the right angle of mandible and extending superior to inferior. Intra-oral examination showed medially displaced right lateral oropharyngeal contents. Ophthalmologic evaluation revealed normal visual acuity with right lateral rectus palsy.

Radiological findings

CT brain done previously was normal. Magnetic resonance imaging was advised for a detailed evaluation, which disclosed a huge 9.5 x 2.6cm lobulated soft tissue mass in the right parapharyngeal region. The mass showed medium signal intensity on T1W1 with strong heterogenous enhancement after contrast. It projected cranially to base of skull via the jugular foramen. The inferior extension was till the level of glottis, as well as extended medially crossing the midline in the retropharyngeal space, posteriorly to prevertebral muscles and supero-laterally below the pinna. It appeared to encase the wall of carotid sheath on right side (Figure 1). There was increased vascularity suggested by multiple small flow voids. MR angiogram was advised which revealed laterally displaced right internal carotid artery with neovascularization in the mass (Figure 2).

Management

A provisional diagnosis of parapharyngeal neoplasm with intracranial extension was made. Systemic examination and hematological

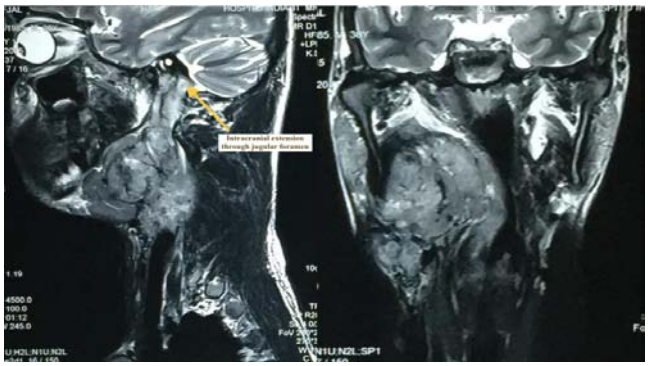

Figure 1: Magnetic resonance imaging: sagittal view showing a huge soft tissue mass in the right parapharyngeal region with cranial extension to base of skull via the jugular foramen and coronal view showing mass in the right parapharyngeal region with inferior extension till the level of glottis and crossing the midline.

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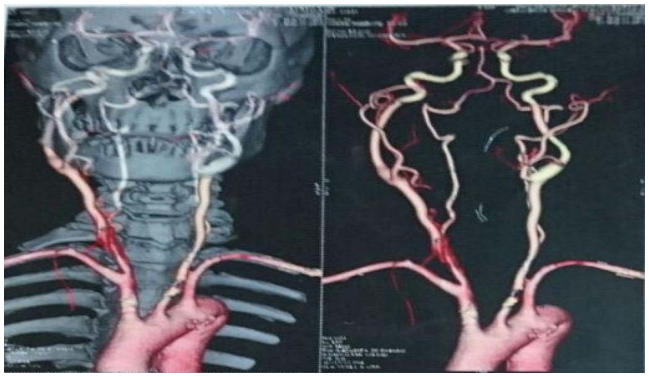

Figure 2: MR angiogram neck showing bowing of internal carotid artery on right side.

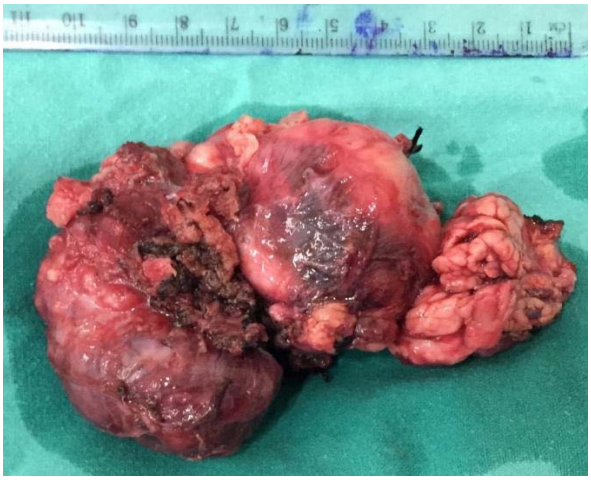

Figure 3: Gross appearance of mass excised in toto by trans-cervical approach.

workup were within normal limits. FNAC was inconclusive. Biopsy was inadvisable due to condition of great vessels. Hence, excision of mass in toto was planned by extended trans-cervical approach. The sternocleidomastoid, posterior belly of the digastrics and stylohyoid muscles were divided, allowing for visualization of the internal and external carotid arteries, internal jugular vein, CN IX, X, XI, XII, and sympathetic chain. Neovascularization was noted and the external carotid artery was ligated for complete clearance. Meticulous resection of all the tumour tissue in toto including intracranial portion extending through jugular foramen was ascertained (Figure 3). Drain was placed and incision was closed in layers followed by pressure dressing. Patient stood the procedure well and discharged on 10th post operative day.

Histopathological Findings revealed grade 2 extra-skeletal mesenchymal chondrosarcoma (Figure 4a) with vascular invasion but no local lymph node metastasis. Immunohistochemistry further confirmed the diagnosis with S-100 (Figure 4b) and CD99 immunoreactive score 3+ and 1+ respectively.

Outcome

Postoperative period was uneventful with adequate healing and good clinical recovery. Later, he underwent postoperative adjuvant

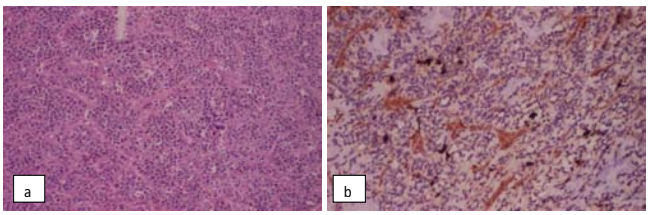

Figure 4: (a) Histopathological examination under 40X magnification revealed round to spindle-shaped cell stroma. (b) Immunohistochemistry S-100 positive (score 3+) under 40X magnification.

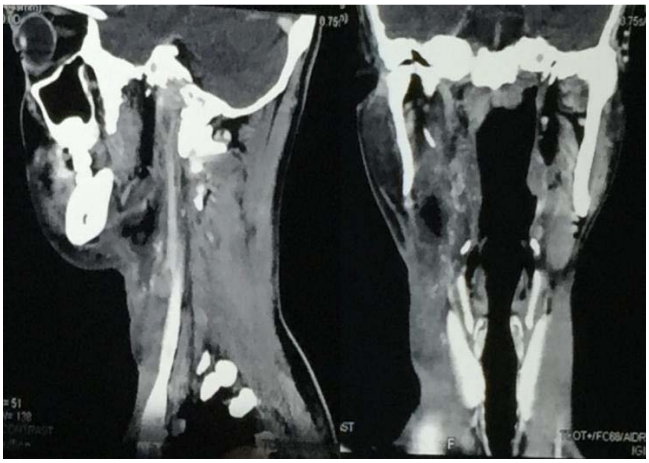

Figure 5: Postoperative computed tomography(sagittal and coronal) showing no obvious residual tumour.

chemotherapy and radiotherapy and was kept under regular follow-up.

There was no obvious residual tumor on postoperative computed tomography done after six months (Figure 5).

Discussion

Primary tumours of the parapharyngeal space are extremely rare, 0.5% of head and neck neoplasms [1] with extra-skeletal mesenchymal chondrosarcoma being exceptionally rare and highly malignant [2]. Only 9 cases of chondrosarcoma were found reported in the parapharyngeal region on searching the available English literature on Pub med including barely one case of extra-skeletal mesenchymal chondrosarcoma published in 1990 [3].

Commonly seen in 2nd decade, it may present as an indistinct neck mass when large enough with intra-oral mild bulging of lateral pharyngeal wall pushing tonsil medially. As a result, it is frequently misdiagnosed as inflammation or tonsil tumour. Thus, its complex anatomical location makes clinical diagnosis very difficult. Even in our case, the patient was initially misdiagnosed with pleomorphic adenoma of submandibular gland.

In the scenario of wavering clinical picture as in our case, imaging studies such as CT, MRI and angiogram are helpful. MRI with gadolinium aids in preoperative assessment of origin, extent and relationship of tumor with surrounding structures and is preferred over CT [1]. It shows a lobulated soft tissue mass iso-intense to skeletal muscle on T1-weighted images and hyper-intense on T2-weighted images that enhance heterogeneously.

Differential diagnosis includes extra-skeletal chondroma and osteosarcoma, synovial sarcoma, hemangiopericytoma, Ewing sarcoma, lymphoma, rhabdomyosarcoma, etc.

It has characteristic histomorphological appearance with two distinct regions of undifferentiated round to spindle-shaped cell stroma and nodules of cartilage and is positive for S-100 and CD99. Biopsy is generally not feasible due to inaccessible anatomical location. However, we or CT-guided FNAC usually determines the nature of the mass, unlike in our case where it proved inconclusive [4].

The treatment comprises of complete surgical resection with adjuvant radiotherapy and chemotherapy. Radical surgery by transcervical approach is the treatment of choice [5,6]. Our case had intracranial extension along with encroachment to great vessels, so we adopted this approach with wide transcervical incision to accomplish adequate exposure and meticulous resection without damaging vital surrounding structures viz. lower cranial nerves namely IX, X, XI, XII, internal carotid artery and internal jugular vein. This prevents possible peri-operative vascular and post-operative neurological morbidities. There was no postoperative mortality or morbidity and our patient is under regular follow-up. Nevertheless, the overall disease prognosis is poor with 5-year survival rate varying from 30 to 50% [5].

Conclusion

Being an extremely rare clinical entity, not much is known about the management of such tumours. Further, its close proximity to the crucial structures in this area warrants a thorough preoperative evaluation for extent and nature of disease and to adopt a careful surgical approach that provides maximum tumor resection with minimum complications and reduces risk of recurrence.

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