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CLINICAL REPORT

A Rare Case of Extraosseous Osteosarcoma (EOS) of Parotid Gland

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Abstract Extraosseous osteosarcoma is a rare malignant soft tissue neoplasm, and extraosseous osteosarcoma of the parotid gland is very rare. It has a very aggressive course, and there are no standardized treatment guidelines. We report the case of a 20 year old male patient who presented with history of right neck swelling since 6 years for which he had undergone right parotid surgery 5 years ago. The final histopathological report indicated that the mass was a pleomorphic adenoma. One year after the first surgery, the patient experienced recurrence of swelling over the operated site, and the size of the swelling has been increasing gradually since then. He was evaluated clinically, and a large mass was noted over the upper aspect of the right upper neck, extending to the occipital and parotid regions. An MRI scan was done which showed a 12 × 10 × 8 cm lesion centred in the right parotid gland, involving paraspinal muscles, C1–C2 vertebrae and extending into the parapharyngeal space. FNAC of the lesion showed features of pleomorphic adenoma. The patient underwent a complete excision of the tumour. The patient's post-operative period was uneventful. The final histopathological report of the patient was extraosseous osteosarcoma of the parotid gland. The patient was referred for adjuvant radiotherapy. He has been on regular follow-up for the past 6 months and has shown no sign of recurrence. EOS is an extremely rare tumour of the head and neck region which often requires extensive surgical resection with or without adjuvant radiotherapy. It has a high rate of local recurrence and a very low disease free survival. Such patients should be kept on a close follow-up.

Keywords Extraskeletal osteosarcoma · Extraosseous osteosarcoma · Parotid gland cancer

Introduction

Osteosarcoma is the most common malignancy of bone, commonly seen in extremities. Only 10% cases of osteosarcomas are diagnosed in the head and neck region [1]. Extraosseous osteosarcoma is a separate term used to describe a very rare and aggressive malignant tumour accounting for less than 1% of soft tissue sarcomas and are extremely rare in the head and neck region [2].

Case report

A 20 year old male patient presented to us with a history of right sided parotid swelling for the past 6 years. He had undergone right superficial parotidectomy/enucleation of mass 5 years ago for the same, the complete surgical details were not available with the patient. The final histopathology report was pleomorphic adenoma. One year after the first surgery the patient had recurrence of swelling over the operated site which has been slowly progressing in size and has reached the current size over the past 3 years.

On clinical examination, a large firm mass was identified over the right parotid measuring about 10 × 8 cm extending to upper neck (levels II–III) inferiorly, anteriorly upto the anterior border of parotid gland, posteriorly to the occipital

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region, superiorly upto the level of tragus, intraorally the mass was seen pushing the tonsillar pillars medially (Fig. 1). Facial nerve functions were intact. Fibreoptic laryngoscopy was done which showed a normal study with mobile vocal cords. Gag reflex of the patient was intact. Examination of all cranial nerves was normal.

A magnetic resonance imaging (MRI) showed $12 \times 10 \times 8$ cm large heterogeneous mass lesion with central cystic, non enhancing areas centred in the right parotid gland involving both superficial and deep lobes, with extension into masseter space and right parapharyngeal space reaching upto C1–C2 vertebrae levels with possible involvement of the same. Right internal carotid artery was seen draping over the medial part of the lesion and was also displaced medially but with preserved signal flow void. Anteriorly the mass was seen abutting pterygoid muscle, posteriorly seen extending into the posterior triangle and right paraspinal muscles, reaching upto midline (Figs. 2, 3 and 4) and caudally it was reaching just below submandibular gland. An FNAC was performed and the report suggested features of pleomorphic adenoma of right parotid gland.

The surgical excision comprised of a total conservative parotidectomy with ligation of external carotid artery and included a wide margin of soft tissue excision which comprised of the muscles of prevertebral, parapharyngeal and retropharyngeal space. Possible lysis was noted over C1–C2 vertebrae levels which was chiselled out after consultation with a spine specialist and excision margins were widened intraoperatively. Facial nerve and all its branches were preserved. (Fig. 5)

Final histopathological examination showed chondroblastic extraosseous osteosarcoma with osteoid and chondroid matrix arranged in an atypical formation. (Fig. 6) Immunohistochemistry was negative for cytokeratin. The postoperative course was uneventful and patient was referred

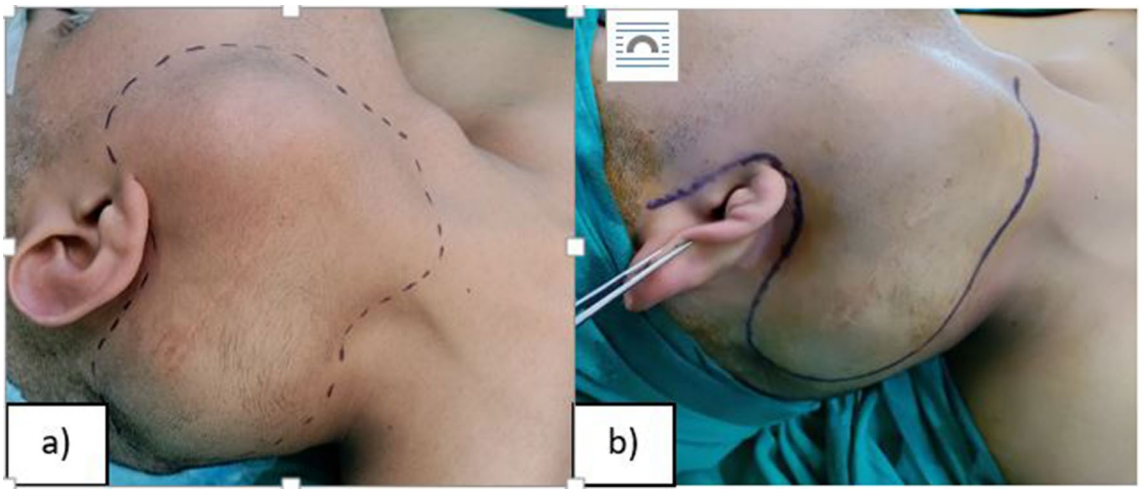

Fig. 1 **a** dotted marker indicating the extent of the mass clinically on lateral view. **b** surgical incision marking

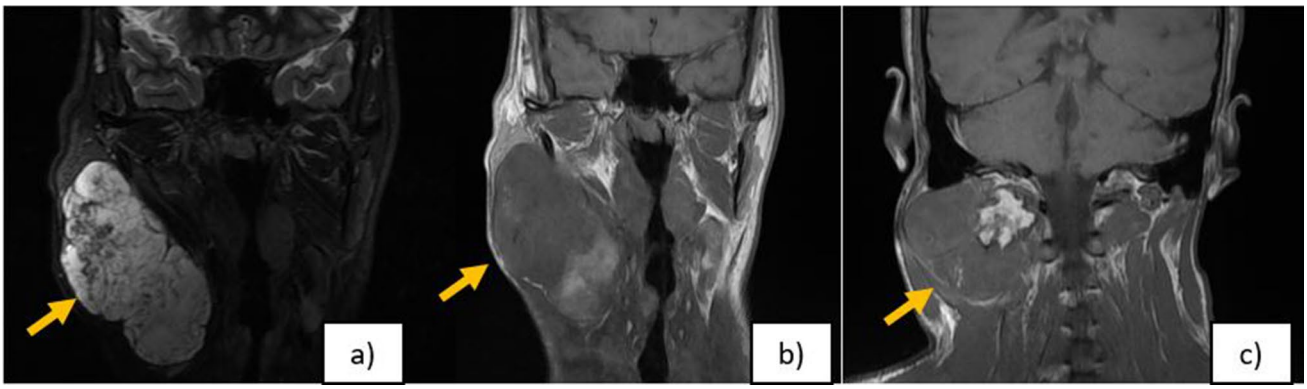

Fig. 2 **a** MRI with contrast axial image. **b, c** MRI plain axial image. Images showing a right parotid mass involving both superficial and deep lobes with extension to C1–C2 vertebral levels

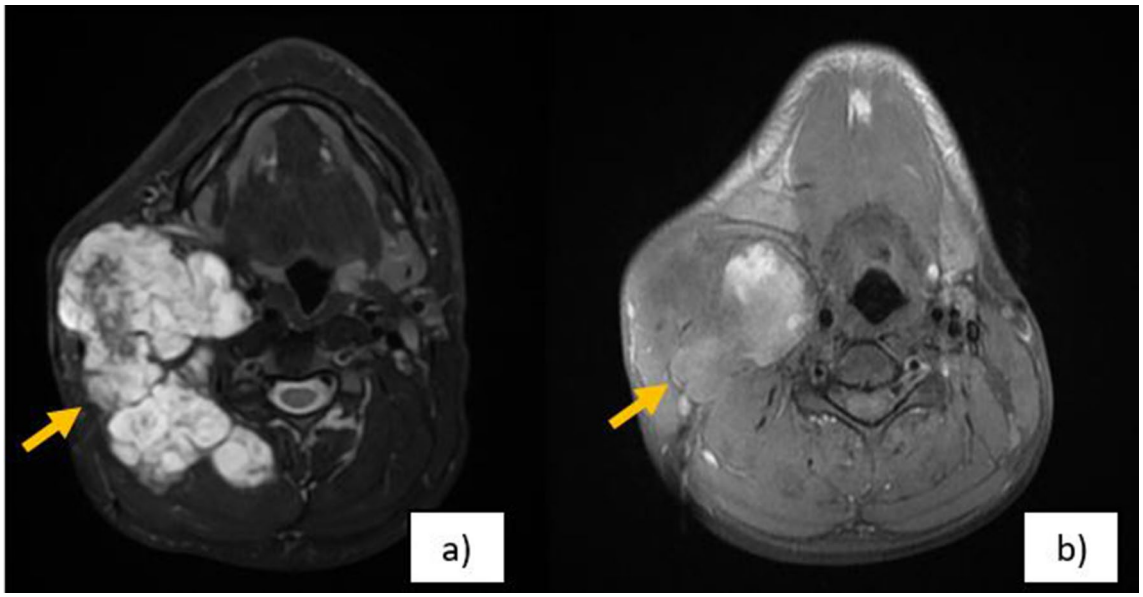

Fig. 3 **a** MRI coronal with contrast. **b** MRI coronal plain. Images showing a right parotid mass involving the Paraspinal muscles, extending to parapharyngeal space

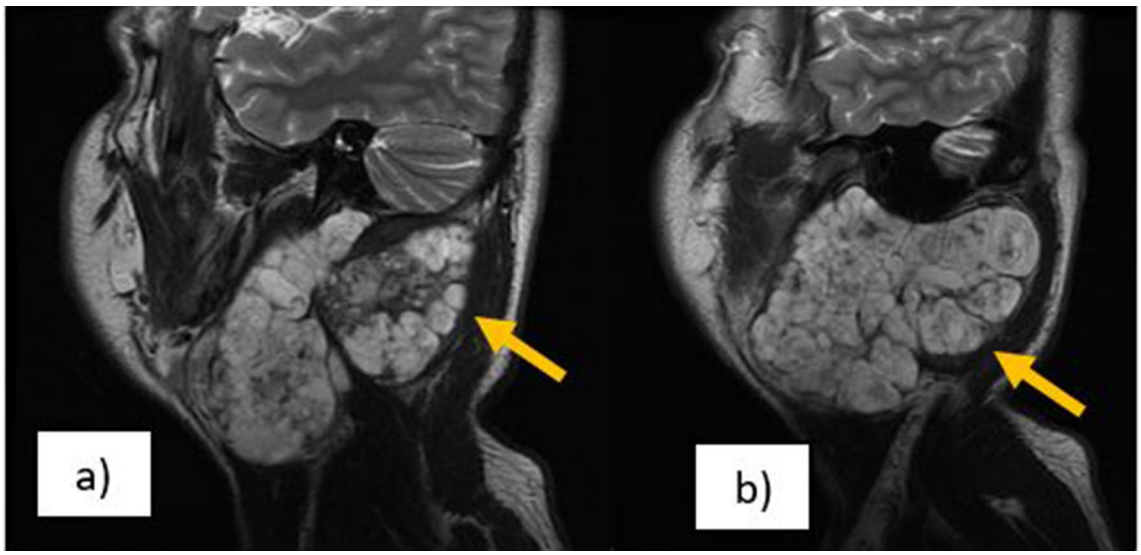

Fig. 4 **a**, **b** MRI sagittal with contrast mass centred in parotid extending to C1–C2 transverse process

for radiotherapy. Follow up scans done after 6 months did not indicate any evidence of residual or recurrent disease.

Discussion

Extraosseous osteosarcoma of the head and neck region is extremely rare and only 15 cases have been reported in available literature out of which 9 cases were seen arising from the parotid gland (Table 1).

The tumour often mimics a reactive lymph node or benign disease as was in the case of our patient. Other differential diagnosis includes myositis ossificans, calcified lymph nodes associated with granulomatous inflammatory disease and pilomatricoma [6]. It has been reported that trauma and radiotherapy are possible inducing agents for extraosseous osteosarcoma [10].

Previous data have shown that FNAC or biopsy cannot produce a reliable diagnosis, with excisional biopsy remaining the gold standard in confirming the diagnosis [14]. Previous reports have also stated that 25% of the cases appeared

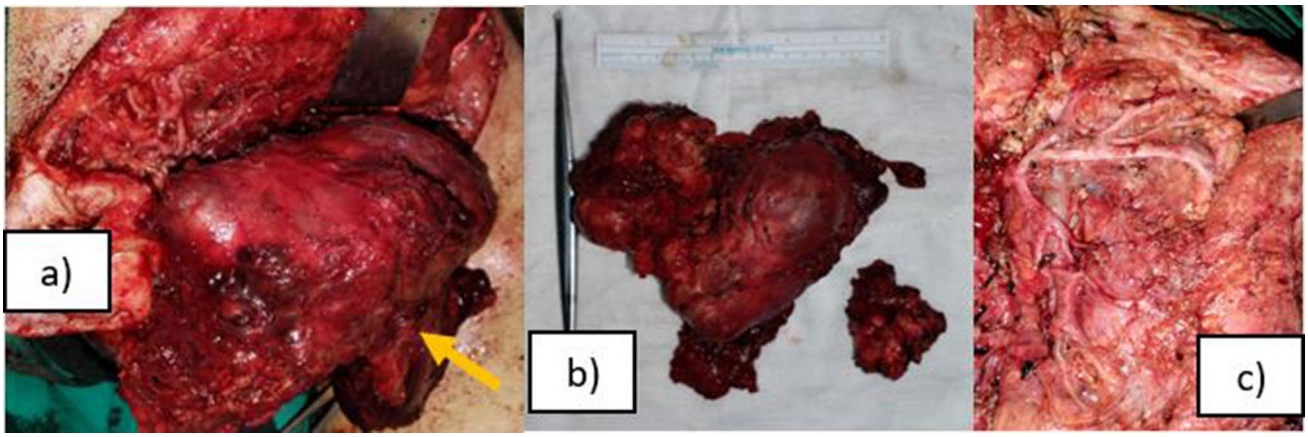

Fig. 5 **a** large mass visualised partly dissected out with the facial nervepartially seen over the bed. **b** 10 × 8 cm well encapsulated mass along with a 4 × 5 cm hard lesion removedseparately. **c** Post operative bed showing all branches of facial nerve intact

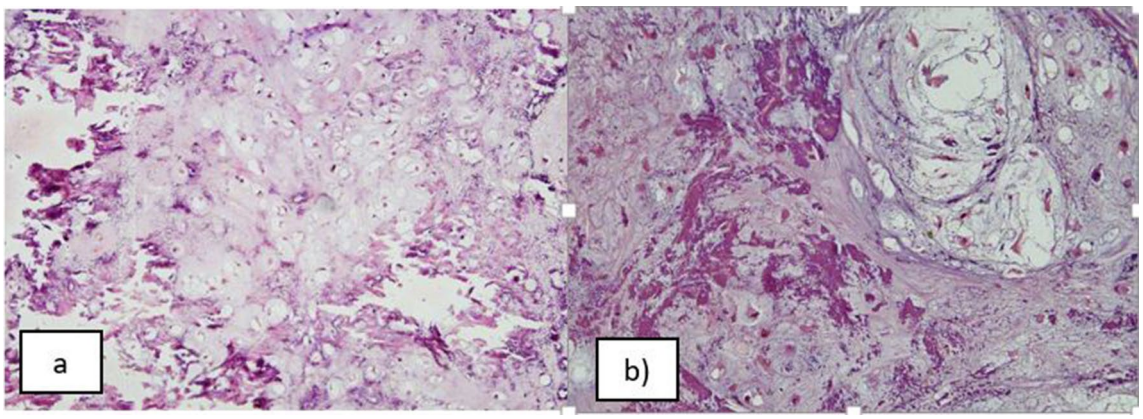

Fig. 6 **a** Haematoxylin and eosin stained section, 100x: shows a tumour composed of lobules of chondrocytic cells within lacunae; left half of the image shows presence of osteoid matrix. **b** Haematoxylin and eosin stained section, 200X: Higher magnification shows chondroid areas containing tumour cells in lacunar spaces that are intimately associated with osteoid matrix (darker pink) present in the left half of the image

encapsulated on gross examination, but on histopathologic examination, all of the tumours penetrated their pseudocapsule and infiltrated the adjacent structures [15]. Histological types include osteoblastic, chondroblastic, fibroblastic, osteoclastic, telangiectatic and small cell forms [3].

Existing data on extraosseous osteosarcoma have emphasized the importance of timely surgical intervention with adjuvant radiotherapy [16]. Recent studies have shown that 88% of reported cases have locally advanced disease (Stage III) according to American Joint Committee on Cancer staging system and 50% cases develop recurrence following surgical resection with wide margins [17]. Tumour diameter of more than 5 cm is an independent poor prognostic factor which directly affects the rate of recurrence in extraosseous osteosarcoma [2]. One report does suggest surgery combined with a chemotherapy (doxorubicin, cisplatin, and ifosfamide etc.) regimen was used to treat EOS, thereby giving a higher survival rate [18]. For parotid EOS, nevertheless, postoperative chemoradiotherapy have not shown any benefits in survival rates till date [10].

The 5 year disease specific survival rates range from 25 to 46% mostly because of the invasive local recurrence and pulmonary metastasis occurring in first post-operative year in 60% patients. Chemotherapy treatment has shown very limited success in 13–15% patients [7]. Due to the rarity of EOS in the head and neck region, it is difficult to obtain a sufficient sample size to evaluate clinicopathological characteristics of this disease and devise evidence-based treatment strategies. Further study is therefore warranted, especially with respect to adjuvant treatment.

Table 1 – Reported cases of extraosseous osteosarcoma in head and neck

	Luna- Ortiz et al. [2]	Manning JT et al. [3]	Hatano et al. [4]	Saito et al. [5]	Stimson et al. [6]	Kosec et al. [7]	Adeyemo et al. [8]	Huang EC et al. [9]	Hamamoto et al. [10]	Xin Ge et al. [11]	Zhang et al. [12]	Aslan et al. [13]	Our case
Age (years)	74	73	25	17	63	48	46	62	47	51	81	69	20
Gender	Female	Male	Male	Male	3 male and 1 female	Female	Male	Male	Male	Male	Male	Male	Male
Localization	Right submandibular region	Parotid gland	Right mandibular angle	Left submandibular region	Parotid	Right neck	Parotid	Parotid	Parotid gland	Parotid gland	Right neck subcutaneous tissue	Parapharyngeal space	Parotid
Tumour diameter (cm)	12	3	1.5	6.5	3–6	6	16	6	6	4	4	5	12
Follow up	Died within 1 year	Lost to follow up	Disease free – 2 years	Died within 1 year	3 died of metastatic disease within 2 years, 1 was lost to follow up	Died of metastatic disease after 1 year	Disease free – 4 months	Died due to metastatic disease in 6 months	Died due to metastatic disease in 17 months	Died due to local recurrence and distant metastasis in 13 months	Disease free – 2 years	Disease free – 6 months	Disease free after 6 months
Treatment	Surgery and palliative radiotherapy	Surgery	Surgery and chemoradiotherapy	Surgery and chemoradiotherapy	Surgery	Surgery	Surgery and chemoradiotherapy	Surgery and chemoradiotherapy	Surgery and radiotherapy	Surgery and chemotherapy	Multiple surgeries with radiotherapy	Surgery and chemotherapy	Surgery and radiotherapy

Conclusion

Extraosseous osteosarcoma is an extremely rare malignancy that encompasses less than 1% of head and neck soft tissue sarcomas. It is difficult to diagnose as it is usually labelled as a benign tumour in its early stages and often requires extensive resection with wide margins. It has a very high rate of local recurrence and very low disease free survival rates.

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Data Availability All material submitted in the article are original.

Declarations

Conflict of interest No potential conflict of interest relevant to this article exist.

Informed Consent Written and informed consent has been taken from the patient for the procedure and all procedures have been done ethically.

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