Intraparotid lymph node with metastatic angiosarcoma: A rare tumor with a routine presentation

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ABSTRACT

Introduction: We present an unusual case presenting as a parotid tumor in a Caucasian man, including our management and a review of literature. Case Report: A 83-year-old male was referred to the ENT department with a non-painful swelling in front of his right ear. This was diagnosed as a likely Warthin’s tumor and this was further supported by ultrasound imaging. Fine needle aspiration cytology was non-diagnostic. The patient underwent superficial parotidectomy for histological confirmation. Histology identified an intraparotid lymph node with metastatic angiosarcoma. The primary source was not identified. The patient was further treated with adjuvant radiotherapy and at a 10-month post-therapy review there is no evidence of reoccurrence. Conclusion: An angiosarcoma is a rare malignant neoplasm of endothelial-type that is characterized by rapidly proliferating and infiltrating anaplastic cells. Angiosarcoma's account for less than 0.1% of all head and neck cancers and it is extremely rare for the major salivary glands to be affected; 80% of all parotid tumors are benign. Metastatic angiosarcoma arising in an intraparotid lymph node has not previously been reported in English journals. This presented case is unusual in its location and its mode of presentation.

Keywords: Intraparotid, Angiosarcoma, Warthin’s, Metastatic

INTRODUCTION

An angiosarcoma is a rare malignant neoplasm of endothelial-type that is characterized by rapidly proliferating and infiltrating anaplastic cells. Angiosarcoma affecting the soft tissues commonly affects the extremities, retroperitoneum and abdominal wall and cutaneous angiosarcoma of the scalp and face is the most common form of angiosarcoma [1, 2]. Although approximately 50% of angiosarcomas occur in the head and neck region, they represent less than 0.1% of all head and neck malignancies and there are very few published cases occurring within the parotid gland [3]. Often these tumors are initially mistaken for other pathology on clinical grounds with diagnosis only made after surgical resection is complete.

CASE REPORT

An 83-year-old male was initially referred to the oral and maxillofacial surgeons with a swelling on the right side of his mouth and in front of his ear. The patient’s complaint was of a painless swelling that was growing over a period of two months. There was no history of discharge in the mouth, weight loss or anorexia. He was referred to the ENT department with an initial diagnosis
of a pleomorphic adenoma. ENT review identified a right parotid swelling and there was no facial nerve palsy (House-Brackmann grade I). Following this, the patient was given a diagnosis of a likely Warthin’s tumor on clinical grounds. His past medical history consisted of two basal cell carcinomas (BCC); one on the chest and one on the right lower leg in the last 6 years. He also suffered from type 2 diabetes, hypertension, and angina.

The patient underwent an ultrasound scan which identified features consistent with a Warthin’s tumor and a fine-needle aspirate (FNA) performed was reported as highly suggestive of neoplasia of an indeterminate type. The patient was consented for a superficial parotidectomy for histological diagnosis and this was performed successfully with full preservation of facial nerve function. The patient was discharged on the first postoperative day and reviewed in clinic with results of the histology. Provisional histology results identified an encapsulated neoplasm with lymphoid tissue present at the edges and thus identifying an intraparotid lymph node. Microscopy identified appearances of an unusual malignant spindle cell neoplasm. Immunohistochemical staining was positive for CD34 and vimentin. The specimen was reviewed at the regional head and neck unit and confirmed a diagnosis of an intraparotid lymph node with metastatic angiosarcoma (Figure 1).

The patient was reviewed by the oncologists and further investigations in the form of staging CT scanning identified no primary site. He underwent 20 doses of adjuvant radiotherapy over a four-week period and a 10-month post-radiotherapy there is no clinical evidence of recurrence.

DISCUSSION

The parotid gland is the most common site for salivary gland tumors of which 80% are benign. The commonest benign tumors are the pleomorphic adenoma (80%), followed by Warthin’s tumor [4]. Malignant parotid tumors are rare with pathologies including mucoepidermoid carcinoma, adenoid cystic carcinoma, adenocarcinoma, malignant degeneration of a pleomorphic adenoma and, squamous cell carcinoma.

Angiosarcomas most commonly occur in skin and soft tissue [2]. They account for less than 0.1% of all head and neck malignancies and it is extremely rare for the major salivary glands to be affected [3]. These tumors are rare malignant neoplasms of endothelial-type cells that line vessel walls; characterized by rapidly proliferating anaplastic cells that infiltrate extensively. Presentation often consists of a moderately paced growing mass associated with compression of adjacent neurovascular structures. There may be systemic features. For example, bleeding and thrombocytopenia, characteristic of a malignant vascular proliferation [5, 6]. This case is unique in that the patient had no other symptoms apart from the swelling occurring over a five month period. On review of literature, we identified seven cases of angiosarcoma involving the parotid gland and no cases of metastatic angiosarcoma arising in an intraparotid lymph node [7, 8, 9].

The diagnosis of a vascular tumor can often be made with the aid of CT or MRI imaging and biopsy in the form of a FNA or core biopsy [10]. Imaging can also aid in the detection of metastases which can occur in up to 50% of cases. Metastases are most commonly seen locally in the regional lymph nodes or distantly in the lungs, liver, or spleen, and sometimes in the both regions.

Studies have shown the 5-year survival of angiosarcoma can vary from 12–41% [5, 6, 11]. The mainstay of therapy is surgical resection with either pre- or postoperative radiotherapy. Tumor site has shown to be prognostically significant in several studies, for example, Morrison et al. demonstrated poor survival in patients with angiosarcoma of the scalp [12]. There is little evidence available demonstrating survival rates in patients presenting with an unknown primary but survival rates have been shown to be associated with early surgical resection prior to local or distant spread [5].

At present there are no clear guidelines on the management of head and neck metastatic angiosarcoma from an unknown primary. Surgical resection and radiotherapy are currently the treatment options with the role of chemotherapy not fully established in the various forms of angiosarcoma [1, 13]. Due to the rarity

![Figure 1: Histology slide of the angiosarcoma at high power using H&E staining.](image-url)
of the pathology, we sought advice from the UK National Sarcoma Unit and our patient underwent adjuvant radiotherapy.

CONCLUSION

Intraparotid angiosarcoma is a very rare pathology which may present through a routine head and neck lump clinic. We present an unusual presentation of such a lesion.

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Author Contributions
Hitens Joshi – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
David McPartlin – Critical revision of the article, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES