An unusual cause of a lump in the parotid gland

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SUMMARY

We present a case of a 50-year-old man with a lump in the left parotid gland. A 4 cm soft mobile lump was identified in the left parotid gland clinically. CT of the parotid glands and neck was arranged confirming a 4 cm mass, predominantly situated within the superficial lobe anterosuperiorly but extending deep to the expected plane of the facial nerve. The positron emission tomography scan did not show distant metastasis. Complete excision of the tumour was achieved. Histological and immunohistochemical findings were consistent with Extraskeletal Ewing's sarcoma. He is undergoing chemoradiation therapy following surgery.

BACKGROUND

Extraskeletal Ewing's sarcoma (EES) is an aggressive malignant tumour of neural crest origin that can develop in the soft tissues and primary EES in the parotid gland is extremely rare.

EES presents a diagnostic dilemma due to its rarity, unclear histogenesis and the heterogeneity of its appearance, requiring a combination of histological, cytological and immunohistochemical analyses for diagnosis. We report the clinical presentation, diagnostic approach and treatment of our patient. This case highlights the diagnostic challenge of EES.

CASE PRESENTATION

A 50-year-old man previously found to be fit was referred to a head and neck surgeon with a painless lump in the left parotid gland of 3 months' duration. The examination revealed a 4 cm well-defined soft mobile mass palpable in the superior area of the left parotid gland. The left facial nerve was functional and the head and neck examination was otherwise unremarkable.

INVESTIGATIONS

CT scans of the parotid glands and neck revealed a 4 cm×3 cm homogeneous ovoid mass, predominantly situated within the superficial lobe of the left parotid gland (figure 1) anterosuperiorly but extending deep to the expected plane of the facial nerve (figure 2). There was a subtle erosive change within the posterior aspect of cortex of the condylar neck. The positron emission tomography scan did not reveal any distant metastasis. Histologically, the specimen from initial fine needle aspiration was composed of loose sheets and singly dispersed monotonous cells with round to oval often eccentric nuclei, finely dispersed chromatin and variably apparent nucleoli. The neoplastic cells had delicate cytoplasm with ill defined cytoplasmic borders and the nuclei were often eccentrically positioned, with many dispersed cells stripped of cytoplasm. In several foci, the neoplastic cells were associated

with delicate blood vessels. The cytological appearances were of an unusual neoplasm but its nature is uncertain. It did not appear to be epithelial or melanocytic and there were no features to suggest a lymphoproliferative disorder. Surgical excision was recommended for definite tumour typing and determination of biological potential. The histology and immunohistochemistry of the surgical specimen showed a mixture of single cells and clusters of loosely cohesive cells. The cells contained round to oval and often eccentrically placed nuclei which were mildly hyperchromatic and variable in size. There was positive staining for PAX B in tumour nuclei and intracellular glycogen deposition. Epithelial markers such as CK-MNF, CK Cam 5.2 and EMA were negative. There was no staining for HEPAR1 (liver marker), PSA (prostate marker), PCC (renal cell carcinoma marker) and S100 (melanoma marker). The tumour cells exhibited CD99, BCL-2 and Vimentin positivity as well as EWSR1 rearrangement. These features were consistent with EES.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses of round cell malignancies affecting soft tissues include primitive neuroectodermal tumours, synovial sarcoma, a poorly differentiated form of solitary fibrous tumour or myoepithelioma. In this case, based on the exhibition of CD99 positivity, BCL-2, Vimentin, EWSR1 rearrangement and cytoplasmic glycogen by the tumour cells, a diagnosis of Ewing's sarcoma of the parotid gland was made.

TREATMENT

Left parotidectomy was carried out. A modified Blair incision was made and the flap was raised. After the facial nerve was identified, the tumour was excised, with surrounding normal parotid gland, and preservation of the facial nerve.

OUTCOME AND FOLLOW-UP

Postoperative recovery was routine and there were no complications. The facial nerve function was normal. After the diagnosis was made, the case was discussed in a multidisciplinary meeting and the decision was made to proceed with adjuvant chemoradiation therapy.

DISCUSSION

Tumours affecting salivary glands are uncommon and most of them are of epithelial origin. Non-epithelial parotid gland tumours are extremely rare with only a handful of cases of mesenchymal tumours affecting parotid glands reported in the literature.^{1–5}

EES is an aggressive neoplasm of neural crest origin, which is histologically similar to primary osseous Ewing's sarcoma. EES with heterogeneity

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Figure 1 Coronal CT image showing a 4 cm×3 cm homogeneous mildly enhancing well-circumscribed ovoid mass situated within the superficial lobe of the left parotid gland.

of its appearance and the similarities shared with primitive neuroectodermal tumours may represent the most undifferentiated end of the spectrum of a group of histogenetically-related tumours known as small round cell tumours.⁶ However, EES differs from primary osseous Ewing's sarcoma in its aggressiveness, distribution and demographics involved. EES usually arises in the soft tissues of the trunk, pelvis and lower extremities. EES in the head and neck regions is rare with cases of EES involving the face, parotid gland, scalp and nasal fossa previously reported.^{1–5 7–11} The average age of occurrence of EES is 20 years compared with 10 years in primary skeletal Ewing's sarcoma. However, the advanced age of our patient is not unprecedented.⁴

The diagnosis of EES is based on the findings of Vimentin, intracellular glycogen deposition, chromosomal translocation t(11:22) along with CD 99 positivity and EWSR1 rearrangement.⁵ The final diagnosis of our case was based on the exhibition of CD99 positivity, BCL-2, Vimentin, EWSR1 rearrangement and cytoplasmic glycogen.

EES usually follows an aggressive course with a high rate of recurrence. Distant metastases are common, most frequently to the lung. Although the prognosis for this tumour is grave with a 5-year survival rate of 20–30% compared with 75% 5-year survival rate of skeletal Ewing's sarcoma, it is reported to be potentially curable.⁴

The treatment of choice is multimodal, just as it is when Ewing's and extraskeletal Ewing's sarcoma strike other sites. Once the diagnosis is established by way of biopsy, surgery followed by chemoradiation therapy may offer the best possible aggressive combination^{4 11} with a study showing that patients

Figure 2 Axial CT scan showing the lesion extending medially posterior to the mandibular ramus to the expected plane of the facial nerve.

treated with surgery, even when the margins were narrow, fared better than those patients who did not undergo tumour resection.⁴

We report an unusual case of EES affecting the parotid gland and highlight the diagnostic challenge of EES. Despite the appreciable rarity of intraparotid extraskeletal Ewing's sarcoma, it is advisable to consider this tumour in the differential diagnosis of a non-calcified soft tissue mass in this region of the body.

Learning points

- ▶ Immunohistochemistry is often indicated if histology of the parotid gland tumour specimen is uncertain and suspected to be of mesenchymal origin.
- ▶ Rapid diagnosis is essential which allows surgical treatment and chemoradiation therapy to take place before metastasis occurs.
- ▶ A multidisciplinary approach to the management of patients with EES in the parotid gland is essential as in the management of all other head and neck cancer cases.

Competing interests None.

Patient consent Obtained.

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