Primary carcinosarcoma of the parotid gland

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Abstract

Carcinosarcoma is a rare malignant ‘mixed’ tumour in the head and neck region. We present a case of carcinosarcoma in a long standing parotid lump and share our experience in the management of the disease together with a review of recent English literature on the subject.

Introduction

Carcinosarcoma is an exceedingly rare malignant mixed tumour in the head and neck region. In the salivary glands, carcinosarcoma comprised only 0.04-0.16% of all malignant salivary tumours1,2 with 65% occurring in the parotid gland.2 These types of tumour have an aggressive characteristic and are often regarded as a high-grade tumour with distant metastasis occurring in 54% of patients. They commonly metastasise to the lung.1 Although, liver, bone and brain involvement has also been reported in the literature,2 these are often rare. Patients usually present between 60-65 years of age3 and most series report no sex predominance.2,4

In this report, we described our experience in managing a patient with carcinosarcoma of the parotid gland and review of published English literature in the management of malignant mixed salivary gland tumour. Cytologic and histologic features including immunohistochemical results are also discussed.

Case Report

A 68-year-old lady presented with a painless lump behind the angle of her left jaw, which she had for years. It had increased in size over the past six months and there were no associated systemic symptoms except for some non-specific weight loss. On clinical examination, there was a 15 mm immobile firm swelling in the area of the parotid tail. The skin overlying the lump was normal in appearance and temperature. Her facial nerve function and the rest of her cranial nerves examinations were normal. There was no palpable cervical lymph node. The rest of her ENT examinations were normal. Her routine laboratory blood analyses consisting of full blood count, inflammatory markers (ESR and C-reactive protein), urea and electrolytes were within normal limits.

A computed tomography (CT) scan revealed a 2 cm diameter lesion in the superficial portion of her left parotid gland. This has a low attenuation centre with an irregular thick enhancing wall. Further evaluation using magnetic resonance imaging (MRI) showed a heterogeneous mass with increase signal on T1-weighted (Figure 1), low signal on T2-weighted images (Figure 2) and enhancement post-gadolinium (Figure 3). There was no evidence of perineural spread or lymphadenopathy. No distant metastases were found. A fine needle aspiration for cytology (FNAC) was performed. The aspirate showed population of large pleomorphic atypical cells exhibiting irregular nuclear contour, large prominent nucleoli and a moderate amount of cytoplasm. Occasional multinucleated giant tumour cells are also seen. The overall appearances are highly suspicious of a carcinoma.

The patient proceeded to have a left superficial parotidectomy. Microscopically, the histological specimen shows a well-circumscribed tumour, which exhibits some variability in appearance. Centrally, there is a hyalinised degenerative area and at the edge of this, there are some small islands and clusters of malignant epithelial cells. These blend into a more spindled area, which has a sarcomatoid appearance with both a distinctive storiform pattern, and focally numerous multinucleated giant tumour cells. There is no evidence of differentiation. This is in keeping with a primary in the parotid gland. Immunohistochemistry shows that this has biphasic characteristic with a definitive epithelial component and a sarcomatous component. Immunohistochemistry analysis with vimentin, p63, S-100 and cytokeratin were positive. These findings were in keeping with carcinosarcoma of the parotid gland.

As the tumour appears to have extended up to the surgical margin of the specimen, the patient proceeded to have a completion radical parotidectomy sacrificing her facial nerve. There was no residual tumour seen in the resected tissue. Post-operatively, she also had radiotherapy treatment comprising of 60Gy in 30 fractions over 6 weeks due to perineural infiltration seen on her initial histology. A gold leaf implant was inserted into her left upper eyelid to treat her lagophthalmos. She declined further facial reanimation surgery and remained disease free at her five-year follow-up.

Discussion

Carcinosarcoma of the parotid gland was first reported by Kirklin et al. in 1951.5 It is defined as a biphasic tumour containing both malignant epithelial and mesenchymal elements.2

The carcinomatous component is usually a poorly differentiated adenocarcinoma, an undifferentiated carcinoma or a squamous cell.2 The sarcomatous component is usually chondrosarcoma.2 In this case immunohistochemistry revealed a biphasic tumour with a definite epithelial component and a sarcomatous component with the appearances of a malignant fibrous histiocytoma.

Carcinosarcoma is known to have arisen from a pleomorphic adenoma, which is the most common neoplasm of major salivary glands though in some cases, this type of tumour appears to have originated de novo. Some authors believe that pleomorphic adenomas and carcinosarcomas share a common stem cell, possibly a myoepithelial cell.1 Malignant transformation of pleomorphic adenoma occurs in around 5-25% of untreated patients and this encompasses three entities - carcinosarcoma ex pleomorphic adenoma, carcinosarcoma and metastasising pleomorphic adenoma. The latter two are exceedingly rare.6 Therefore, the differential diagnoses of a unilateral parotid swelling in this case include pleomorphic adenoma, haemangiomia, spindle cell squamous carcinoma, primary salivary sarcomas, carcinoma ex pleomorphic adenoma and mucoepidermoid carcinoma. Distant metastasis is usually blood borne.

The diagnosis is made based on a combination of clinical history, physical examination, pathology, and imaging. Cytologically, the
tumour cells display features of atypia, including cellular and nuclear pleomorphism, hyperchromatism, mitotic figures and invasive growth. These features often aid to distinguish carcinosarcomas from benign mixed tumour.7

In this case, several FNAC performed were reported as suspicious and it was not until after the initial parotidectomy that carcinosarcoma was formally diagnosed. Immunohistochemistry such as cytokeratin AE1/AE3, vimentin, S-100 and anti smooth muscle actin can be helpful in differentiating carcinomatous and sarcomatous components.8 Radiological imaging is useful in the planning of the operation and when consenting patient for surgery. CT enables assessment of tumour site and whether the deep lobe of the parotid gland is involved. MRI scans, however, a more useful radiological investigation in assessing for soft tissue and perineural spread. This will also aid in differentiating a malignant tumour from a benign tumour.

The management of patient with carcinosarcoma of the parotid gland is radical surgery.9 Radical parotidectomy usually requires sacrifice of the facial nerve. Therefore, subsequent measures have to be taken to protect the affected eye from corneal ulceration and referring the patient for facial reanimation surgery. Neck dissection is appropriate in those with lymphadenopathy. Although our patient received post-op radiotherapy, there is no substantive evidence at present to support the use of post-operative radiotherapy treatment in increasing survival rates in these patients. However, fast neutron radiotherapy had been used as sole therapy in patients with unresectable salivary gland tumours and this has shown high potential local-regional control and survival rates.10

The prognosis often depends on the clinical and histological stage of the disease. It is difficult to predict the long-term prognosis of primary carcinosarcoma of the parotid gland due to the low prevalence of this disease. The prognosis is generally poor with 58-85% of patients dying of the disease.3,11,12 In a study by Livolsi et al., out of forty-seven cases of malignant mixed tumour, 38% showed local recurrences and a five-year survival of 85% in their case series.13 A population study conducted by Modulin et al. consisting of 13,715 patients with carcinoid tumours over five decades found that the five year survival, regardless of primary site, was only 67.2%.14 Our patient remained free of recurrence five years following her initial presentation and this is probably because she was diagnosed and treated early.

The future in understanding the pathogenesis of carcinosarcoma and to treat this tumour effectively will depend on advancement made in molecular genetics to improve our understanding of the clonal origin of this tumour and the use of improved immunohistochemistry to stage the disease. Gotte et al. concluded from their study favouring the hypothesis of monoclonal origin with a possibility of an inactivated tumour suppressor gene on Chromosome 17 and a wild-type allele of the p53 tumour suppressor gene.15 In recent years, Xin et al. found that Ki67 is a useful staging marker and therefore may have a role in predicting the prognosis of the disease.16

Conclusions

Carcinosarcoma is a rare case of a mixed malignant tumour of the parotid gland. The main stay of treatment is surgical resection. Although our patient received post-op radiotherapy, there is no convincing evidence at present to support the use of post-operative radiotherapy treatment to increase survival in these patients. This is due to the limited number of case series. It is vital that these patients are closely follow-up and observed for symptoms and signs of local recurrence and metastatic disease. Early recognition and intervention are critical for favourable outcomes.

References

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