Acinic Cell Carcinoma of the Parotid Gland in Children

A Case Report and Literature Review

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ABSTRACT

Parotid acinic cell carcinoma is a rare malignancy in childhood. We report the case of a 12-year old girl presenting with a palpable mass in the left maxillofacial area. The radiologic evaluation showed a parotid mass. Tumour resection revealed acinic cell carcinoma of the parotid gland. She underwent complementary total parotidectomy without any adjuvant treatment. The patient has been disease-free for the last five years. We review the literature on acinic cell carcinomas of parotid glands in childhood.

Carcinoma de Células Acinosas de la Glándula Parótida en Niños

Reporte de un Caso y Revisión de la Literatura

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RESUMEN

El carcinoma de células acinosas de la parótida es una malignidad rara en la niñez. Reportamos el caso de una niña de 12 años con una masa palpable en el área maxilofacial izquierda. La evaluación radiológica mostró una masa parótida. La resección del tumor reveló un carcinoma celular de la glándula parótida. Fue sometida a una parotidectomía total complementaria sin tratamiento adyuvante alguno. La paciente ha estado libre de enfermedad durante los últimos cinco años. Revisamos la literatura sobre carcinomas de células acinosas en las glándulas parótidas en niños.

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INTRODUCTION

Salivary neoplasms are uncommon in childhood (1). Among them, malignant tumours are met more frequently in the parotid gland and in older children (2). Due to their rarity, a high index of suspicion is required for diagnosis, and the experience of individual surgeons remains limited. We present a case of acinic cell carcinoma of the parotid gland in a 12-year old girl and discuss the literature.

Case report

A 12-year old girl presented with a painless swelling over the left parotid area. Her physical, laboratory and radiological assessment were otherwise unremarkable. On imaging, the mass seemed to be well defined and was removed under general anaesthesia. On microscopy (Fig. 1), the tumour was composed predominantly of solid sheets of basophilic granular cells. With the exception of some vacuolated cells, the overall tumour appearance was monomorphic and its edge was characterized by prominent lymphoid aggregates. Based on histology and immunohistochemistry (Fig. 2), the pathologist diagnosed acinic cell carcinoma of the parotid gland.

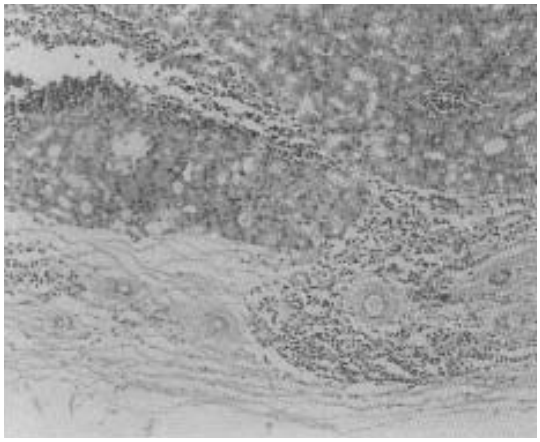

Fig.1: Clusters of neoplastic cells infiltrating the adjacent salivary gland. Haematoxylin-eosin.

A thorough radiologic investigation for secondary disease was negative. The patient underwent complementary total parotidectomy with preservation of the facial nerve.

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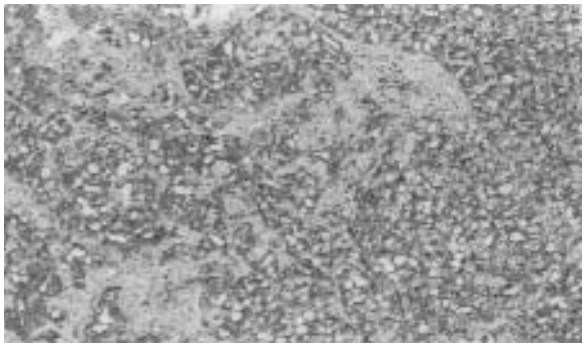

Fig. 2: Immunohistochemical expression of cytokeratin 7 in the neoplastic cells. Streptovidin-Biotin-HRP.

Her postoperative course was uneventful and she remains in good health five years later without evidence of residual or recurrent disease.

DISCUSSION

Childhood salivary gland tumours account for less than 8% of all paediatric head and neck tumours (3) and less than 5% of all salivary gland neoplasms (1). The commonest salivary gland lesions in children are as a result of benign tumours and inflammatory situations (4, 5).

They are mostly localized in the parotid glands, with submandibular localization being much less common (6). Malignancy is met in 16%–50% (5, 7) of parotid tumours in childhood and drops to 15%–25% in adults (8). Epithelial solid salivary tumours in the population usually present between 8 and 20 years of age (3). Average age at surgery is 15.4 years for a benign lesion and 10.5 years for malignancy (7).

Acinic cell carcinomas seem to account for 6–37% of total parotid malignancies in children (2, 5, 7, 9, 10). They are extremely rare under the age of 16 years (11). A female predominance exists (3) although this has not been supported by all series (7). Their pathogenesis is obscure and is probably irrelevant to Epstein-Barr virus infection (12). A single report of familial occurrence exists in the literature (13) but there is no evidence of a genetic trait. Parotid acinic cell carcinomas may recur after chemotherapy or radiotherapy (14).

The primary lesion is usually solitary and well defined. In contrast, recurrences tend to be multinodular with incomplete encapsulation. On microscopy, the tumour has a benign monomorphic appearance with sparse, well-vascularized stroma and may contain lymphoid tissue. The cells have abundant granular cytoplasm resembling serous acini. Clear cells, vacuolated duct cells and vacuolated atypical glandular epithelium can be seen among the glandular cells. Pure clear-cell type acinic carcinomas also exist (11). Ultrastructural, histochemical and morphological studies suggest that the cell of origin is the intercalated duct cell (11, 15).

Acinic cell tumours belong to the solid parotid gland tumours with varying degrees of malignancy (16). They are classified according to their malignant potential. Low-grade malignancies (Grade I) are completely encapsulated without capsule invasion. Moderately malignant ones (Grade II) show signs of capsular invasion. Grade III (high-grade) tumours infiltrate the surrounding tissues and present papillary-cystic zones (17). Adjacent tissue invasion, regional lymphatic dissemination and distant metastasis (lungs and bone) are possible (11, 15, 17, 18).

Acinic cell tumours are painless and slowly growing in most cases (11). Rarely, it presents with features suggestive of malignancy: pain, rapid growth, facial nerve paresis or lymphadenopathy (2, 3, 17, 19).

Physical examination is usually non-informative (10). Imaging may increase suspicion but is also non-diagnostic because of low specificity (20–22). Computed tomography and magnetic resonance imaging can detect infiltration of adjacent structures in high grade malignancies (23). Fine needle aspiration (FNA) has a sensitivity of 82% to 91% and a specificity of 86% to 96% (24, 25) but it can also be misleading (26). Furthermore, its applicability is limited by children's tolerance of interventional examinations (7, 10). The diagnosis can be set by ruling out inflammatory situations and excision biopsy. However, this strategy has been debated because of the possibility of facial nerve damage, tumour spillage and recurrence (10, 26). The recommended treatment of acinic cell carcinoma of the parotid is total parotidectomy with preservation of the facial nerve (7, 9, 10, 26). Infiltration of the nerve or perineural structures may demand its sacrifice and the deficits can be restored with nerve grafting (7, 10, 26). Neck dissection is best reserved for patients with regional lymphadenopathy (10) although some advocate its routine practice (18). The results of radiotherapy are inconsistent and the possibility of probable long-term sequelae cannot be overlooked. Irradiation seems therefore most appropriate in locally advanced disease or in high grade tumours (5, 18, 19). Operative morbidity includes Frey's syndrome and facial numbness (11). Facial weakness due to nerve injury, nerve sacrifice or unrelated to the nerve is unusual (5). Prognosis of acinic cell parotid gland tumours in children is generally good. Five-year survival rates range from 89% to 96% but fall to 56% at 20 years (18). Therefore, surveillance must continue for a long time.

In conclusion, acinic cell parotid carcinoma in childhood is a rare entity that requires a high index of clinical suspicion, prompt diagnosis and definitive initial surgical treatment. If the condition is appropriately dealt with from the beginning, its morbidity is extremely low and permanent cure is highly probable.

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