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Case Report

Pediatric Parotid Acinic Cell Carcinoma Mimicking a Benign Lesion: A Case Report

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Abstract

Acinic cell carcinoma (ACC) is a rare malignant epithelial tumor of the salivary glands, accounting for only 2-5% of all primary parotid neoplasms. Although typically seen in adults with a slight female predominance, its occurrence in children is extremely rare. ACC is considered a low-grade malignancy, often displaying well-differentiated histological features that resemble normal acinar cells of the salivary gland. Radiologically, these tumors often appear well-circumscribed and encapsulated, which can mimic benign lesions such as pleomorphic adenomas, making preoperative diagnosis challenging. In pediatric patients, this resemblance may delay suspicion of malignancy. Therefore, histopathological examination remains essential for a definitive diagnosis and for guiding appropriate management.

Keywords: *Acinic Cell Carcinoma; Pediatric Parotid Tumor; Salivary Gland Neoplasm*

Introduction

Acinic cell carcinoma (ACC) is a rare malignant epithelial tumor of the salivary glands, most commonly arising in the parotid gland and accounting for only 2-5% of parotid neoplasms [1]. It typically affects adults and is exceptionally rare in children. Despite its malignant nature, ACC often appears well-circumscribed and encapsulated on imaging, closely mimicking benign lesions such as pleomorphic adenomas. This imaging overlap can lead to diagnostic challenges, particularly in pediatric patients where malignancy is less expected. We report the case of a 14-year-old child with a parotid mass initially interpreted as benign on ultrasound and MRI, but ultimately diagnosed as low-grade ACC on histopathological analysis. This case underscores the limitations of imaging in reliably distinguishing benign from malignant parotid tumors in children.

Case Report

A 14-year-old girl presented with a gradually enlarging, painless facial swelling. The clinical examination identified a mobile, ovoid swelling located in the right preauricular area. The swelling was soft, non-tender, and without any signs of inflammation. An MRI was first made, which showed a parotidean cystic mass, that exhibits a restricted diffusion with low ADC, without contrast enhancement (Figure 2). Ultrasound imaging identified a hypoechoic cystic lesion within the right parotid gland, with no evidence of vascularity, with dorsal acoustic enhancement and bright internal echoes (Figure 1).

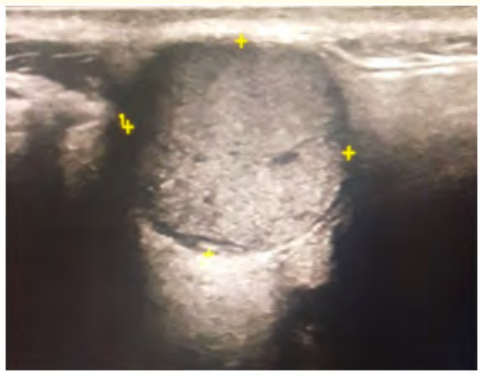

Figure 1: *Right parotid ultrasound showing a well-circumscribed cystic mass with regular margins, posterior acoustic enhancement, and finely echogenic content. No color Doppler signal was detected within the lesion.*

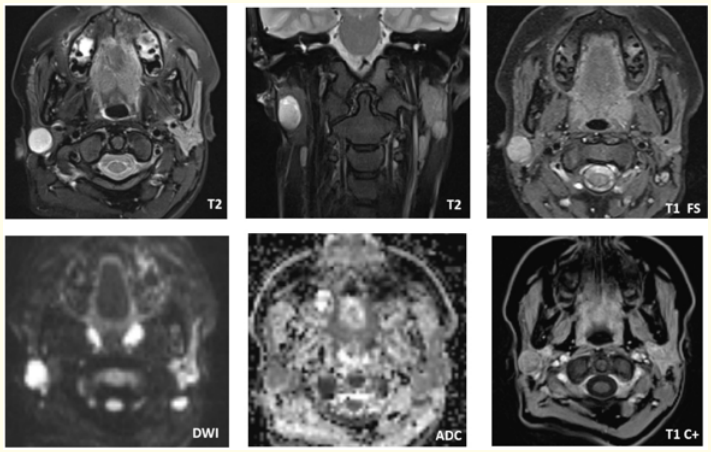

Figure 2: *Parotid MRI showing a well-circumscribed right parotid mass, hyperintense on T2 and T2 FAT-SAT sequences, hypointense on T1, with diffusion hypersignal and slightly low ADC values (0.9 to 1 × 10-3 mm2/s). Mild peripheral enhancement was observed after gadolinium administration.*

Discussion

Acinic cell carcinoma (AcCC) is a rare, typically low-grade malignant salivary gland tumor that most commonly affects the parotid gland [1]. Although once considered benign, it is now recognized for its potential to recur locally and metastasize to regional lymph nodes or distant organs, such as lungs and bones, which necessitates prolonged follow-up [2]. In pediatric patients, AcCC accounts for approximately 3-4% of parotid tumors and is the second most frequent malignant parotid neoplasm after mucoepidermoid carcinoma [3]. Despite limited data in children, the prognosis is generally favorable, with high survival rates, but long-term surveillance is crucial due to possible late recurrences.

Clinically, AcCC presents as a slow-growing, painless mass in the parotid region; however, symptoms such as facial nerve palsy, pain, or rapid growth may indicate more aggressive disease [4].

Radiologically, acinic cell carcinoma of the parotid gland presents no specific or pathognomonic features on CT, MRI, or ultrasound, making diagnosis by imaging alone challenging due to its similarity to benign tumors [5]. Ultrasound is useful for assessing tumor size, location, and characteristics and is commonly used to guide fine-needle aspiration biopsies. Contrast-enhanced CT typically shows mild enhancement with possible hypoattenuating areas indicating central necrosis, alongside an irregular solid component [6]. MRI signals are variable and nonspecific: the tumor may appear well-marginated with a mildly T2 hyperintense solid component and cystic or necrotic areas that are also hyperintense on T2-weighted images. These MRI signals correlate with histological features such as vascularity, fibrosis, hemosiderin deposits, and calcifications [5]. Features raising suspicion of malignancy include regional lymphadenopathy, rapid tumor growth, and clinical signs such as pain. In our case, both ultrasound and MRI showed features suggestive of a benign lesion, illustrating the difficulty of distinguishing ACC from benign tumors based on imaging alone.

Histologically, ACC is a well-circumscribed tumor with variable patterns (solid, cystic, microcystic, follicular, papillary-cystic) and diverse cell types. It typically forms large lobules with little stroma. Histologic grade is debated and poorly correlates with clinical behavior [7]. Although histologic grading remains controversial, histopathology plays a major role in diagnosis, as in our case.

The mainstay treatment for acinic cell carcinoma is complete surgical excision, performed via intraoral or extraoral approach depending on tumor size and location [7]. In our case, the patient underwent a radical surgery without complications, and no recurrence has been observed to date.

Conclusion

Acinic cell carcinoma is an exceptionally rare salivary gland tumor in pediatric patients. Its benign imaging appearance poses a diagnostic challenge, making histopathology essential for accurate diagnosis and treatment planning. As in our case, a combined radiologic and histologic approach enables appropriate surgical management and favorable outcome

Ethical Approval and Patient Consent

Written informed consent was obtained from the patient's legal guardian for publication of this case report and accompanying images.

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Author Contributions

All authors contributed equally to the conception, drafting, and final approval of the manuscript.

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