

Low-grade mucoepidermoid carcinoma of the parotid gland in a 5-year-old child: The youngest reported case from India

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ABSTRACT

Mucoepidermoid carcinoma (MEC) of the parotid gland is scarce in the pediatric group and is exceedingly uncommon in children under 6 years. In spite of being the most frequent malignant salivary gland tumor in children, the majority of cases are reported in older age groups. Only a few cases in children under 5 years have been documented worldwide, and the youngest of these previously reported Indian cases was a 6-year-old child. We report a rare case of low-grade MEC of the parotid gland in a 5-year-old boy, describing the youngest reported case from India to date. The presenting complaint was a painless parotid swelling, and pre-operative imaging and fine-needle aspiration cytology (FNAC) were suggestive of pleomorphic adenoma. FNAC has limited sensitivity in pediatric salivary gland tumors, particularly in low-grade MEC, due to superimposing cytomorphological features with benign lesions. The management comprised superficial parotidectomy with facial nerve preservation, with histopathological examination confirming low-grade MEC with clear margins. This case highlights the rarity of this condition, diagnostic pitfalls and limitations of FNAC, and the significance of histopathological confirmation.

Key words: Mucoepidermoid carcinoma, Pediatric, Parotid, Pleomorphic adenoma

Salivary gland tumors are scarce in children, accounting for a small fraction of pediatric head and neck neoplasms, with parotid tumors being the commonest among them, albeit most are benign, and malignancies are unusual, particularly in early childhood [1,2]. Mucoepidermoid carcinoma (MEC) is the most frequent malignant salivary gland tumor in children, but it typically occurs in older children and adolescents [1-3]. MEC in children under 6 years is exceedingly rare and often presents as a slow-growing, painless parotid mass that can mimic benign lesions such as pleomorphic adenoma [3-5]. Pre-operative fine-needle aspiration cytology (FNAC), while widely used, may be unreliable in low-grade MEC due to overlapping cytological features, resulting in misdiagnosis [6].

Reports of pediatric parotid MEC from India are extremely sparse; we report a rare case of low-grade MEC in a 5-year-old boy, representing the youngest case from India, and highlighting the diagnostic challenges and management considerations [4,7]. Reporting this case contributes to the limited global literature on salivary gland malignancies in very young children. Since this case involves a 5-year-old child, it represents one of the

youngest reported cases from India, adding valuable epidemiological data.

CASE REPORT

A 5-year-old boy reported in our outpatient department with a gradually progressive, painless swelling over the right parotid region for the past 3 months, with no history of fever, trauma, facial asymmetry, or systemic symptoms.

On examination, the child was conscious, alert, and hemodynamically stable. Vital signs were within normal limits: pulse rate 98 beats/min, blood pressure 110/68 mmHg, respiratory rate 20 breaths/min, temperature 36.8°C, and oxygen saturation 99% on room air. Local examination revealed a well-defined, firm, non-tender swelling measuring 3 × 3 cm. The overlying skin was normal, facial nerve functions were intact, and there was no cervical lymphadenopathy.

Pre-operative ultrasonography showed a well-defined, homogeneous hypoechoic lesion in the superficial lobe measuring 1.3 × 1.9 × 1.5 cm, suggestive of a benign lesion. FNAC reported a pleomorphic adenoma (Fig. 1).

The child underwent a right superficial parotidectomy under general anesthesia with

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meticulous preservation of the facial nerve. A modified Blair (lazy-S) incision was made extending from the preauricular region and curving below the ear lobule into the upper cervical region. The parotid fascia was incised, and careful dissection was carried out to identify the main trunk of the facial nerve using standard anatomical landmarks such as the tragal pointer, posterior belly of the digastric muscle, and the tympanomastoid suture. Intraoperatively, the tumor was nodular, and once the facial nerve trunk was identified, its branches were meticulously dissected and preserved. Hemostasis was secured, and the surgical field was irrigated. A closed suction drain was placed in the operative bed, and the wound was closed in layers (Fig. 2).

Grossly, the lesion measured $3 \times 2 \times 1.5$ cm. Histopathology showed mucous, epidermoid, and intermediate cells arranged in cystic and solid patterns, consistent with low-grade MEC. Immunohistochemistry (IHC) confirmed the diagnosis, and surgical margins were free of tumor (Fig. 3).

The post-operative period was uneventful, and facial nerve function was preserved. No adjuvant therapy was

given. The patient remains disease-free at 6 months of follow-up.

DISCUSSION

MEC is rare in children, representing a small proportion of pediatric head and neck tumors, and most cases occur in the second decade [1,2]. Cases under 10 years are unusual, and those under 5 are extremely rare [3-5]. Literature reports include a 4-year-old, a 20-month-old toddler, and a 6-year-old girl from India, making this 5-year-old boy one of the youngest globally and the youngest reported from India [4,7].

Clinically, MEC presents as a painless, progressively enlarging parotid mass; imaging usually shows a solid lesion, but FNAC may be misleading [3,6]. Although FNAC is minimally invasive with high specificity, the sensitivity for MEC is variable, especially for low-grade or cystic tumors [6]. False negatives are common, with over 50% of MEC cases misdiagnosed or underdiagnosed on cytology alone, due to inadequate sampling of cystic areas, low cellularity, or overlapping features with benign lesions [6]. Non-diagnostic FNAC may occur with insufficient representative material. A high clinical suspicion is essential, and definitive diagnosis requires histopathology with IHC [6,8].

Histologically, MEC is characterized by mucous, intermediate, and epidermoid cells in cystic and solid patterns; this architecture may be missed on cytology [1,8]. IHC confirms diagnosis and distinguishes MEC from other parotid tumors; molecular markers such as Mastermind-like transcriptional coactivator 2 rearrangement may further support diagnosis [7-9].

Management principles in children mirror those of adults: complete surgical excision is the cornerstone. Low-grade MEC is best treated with superficial



Figure 1: Pre-operative ultrasonography showing hypoechoic lesion in the right parotid region



Figure 2: Intraoperative image showing facial nerve preservation during parotidectomy

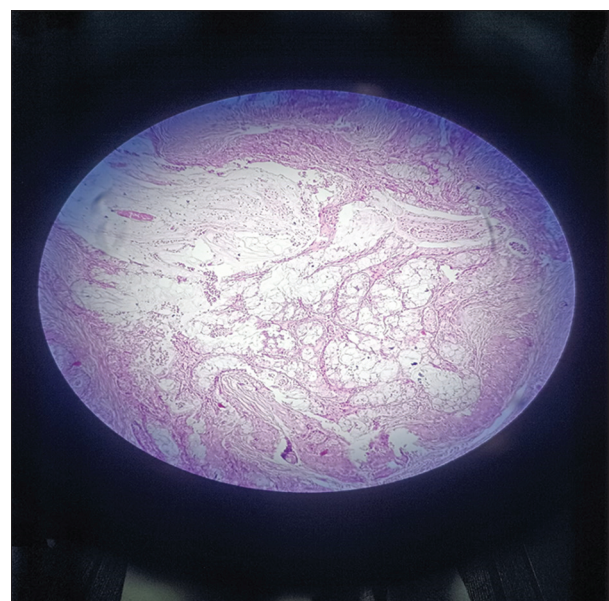


Figure 3: Histopathological image showing mucous, epidermoid, and intermediate cells of mucoepidermoid carcinoma

parotidectomy and meticulous facial nerve preservation to maintain an excellent prognosis and quality of life. Literature shows surgical excision provides high disease-specific survival (>90%) when margins are clear [3,10].

Neck dissection is reserved for clinically positive nodes or high-grade tumors due to the low incidence of occult metastasis in low-grade MEC [3]. Adjuvant radiotherapy is rarely used in children because of long-term side effects and is considered only for high-grade tumors, positive or close margins not amenable to re-excision, or perineural invasion [3,10].

Histopathological examination remains the gold standard for diagnosis and grading, particularly when pre-operative tools are misleading [1,6]. Long-term follow-up is essential due to the possibility of late recurrence, even in low-grade tumors [3]. This case underscores the limitations of FNAC, the value of histopathology with IHC for definitive diagnosis, and the efficacy of surgical management with facial nerve preservation in very young children [3,6,10].

CONCLUSION

MEC in very young children is exceptionally rare. FNAC alone may underestimate malignancy; definitive histopathology with IHC is crucial. Surgical excision with clear margins is the mainstay, and long-term surveillance is important for early detection of recurrence.

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