

Introduction

Salivary glands consist of parotid, submandibular, sublingual & other minor salivary glands. Tumors affecting these glands present a diagnostic and therapeutic challenge. They rare in children, with an annual incidence of 0.08 per 100 000.^[2] When present, older children are affected.^[2] Mucoepidermoid carcinoma (MEC) is the most common cancer presenting in children with a predilection for the parotid gland.^[3] This case report describes presentation of this rare tumor in a three-year old girl with a brief review of the literature.

Case Report

A three-year old female presented to the ENT department at Chris Hani Baragwanath Academic Hospital (CHBAH) with a six-month history of a progressively enlarging, intermittently painful mass at the angle of the jaw. No facial weakness was reported. There was no other significant history on further enquiry.

Clinical examination showed a well-child with normal vital signs. Head & neck examination showed a right sided firm, non-tender mobile parotid mass of approximately 3cm in diameter with normal overlying skin. The mass was confined to the parotid space with no intraoral extension and no lymph nodes were palpable. Facial nerve was intact. Initial assessment was that of a slow growing benign mass, possibly infective or granulomatous, to exclude tuberculosis (TB) being common in our setting.

Contrasted Computed Tomography (CT) scan showed a contained, enhancing, irregular parotid gland lesion (Fig.1). Fine needle aspiration and cytology (FNAC) of the mass was inconclusive. Blood tests, including Human Immunodeficiency Virus (HIV) and Epstein Barr Virus (EBV) were normal. Chest X-ray was normal.

The child underwent superficial parotidectomy via a modified Blair incision, with facial nerve preservation (Fig 2). The procedure was done under general anaesthesia with facial nerve intraoperative monitoring. Perioperative intravenous Augmentin antibiotic was given & continued for 48 hours until the drain was removed. The child was discharged 3 days following the procedure with no facial nerve paresis. The specimen was sent for histopathological evaluation, which showed intermediate grade MEC with uninvolved surgical margins. The tumour was stage III (pT3 pN0 M0). The child was not given postoperative chemotherapy or radiation in view of negative surgical margins & the intermediate nature of the lesion. No recurrence has been observed in 15 months since the procedure. Regular reviews will be continued on a long-term basis.



Fig 2: Exposure of the parotid lesion via a modified Blair incision

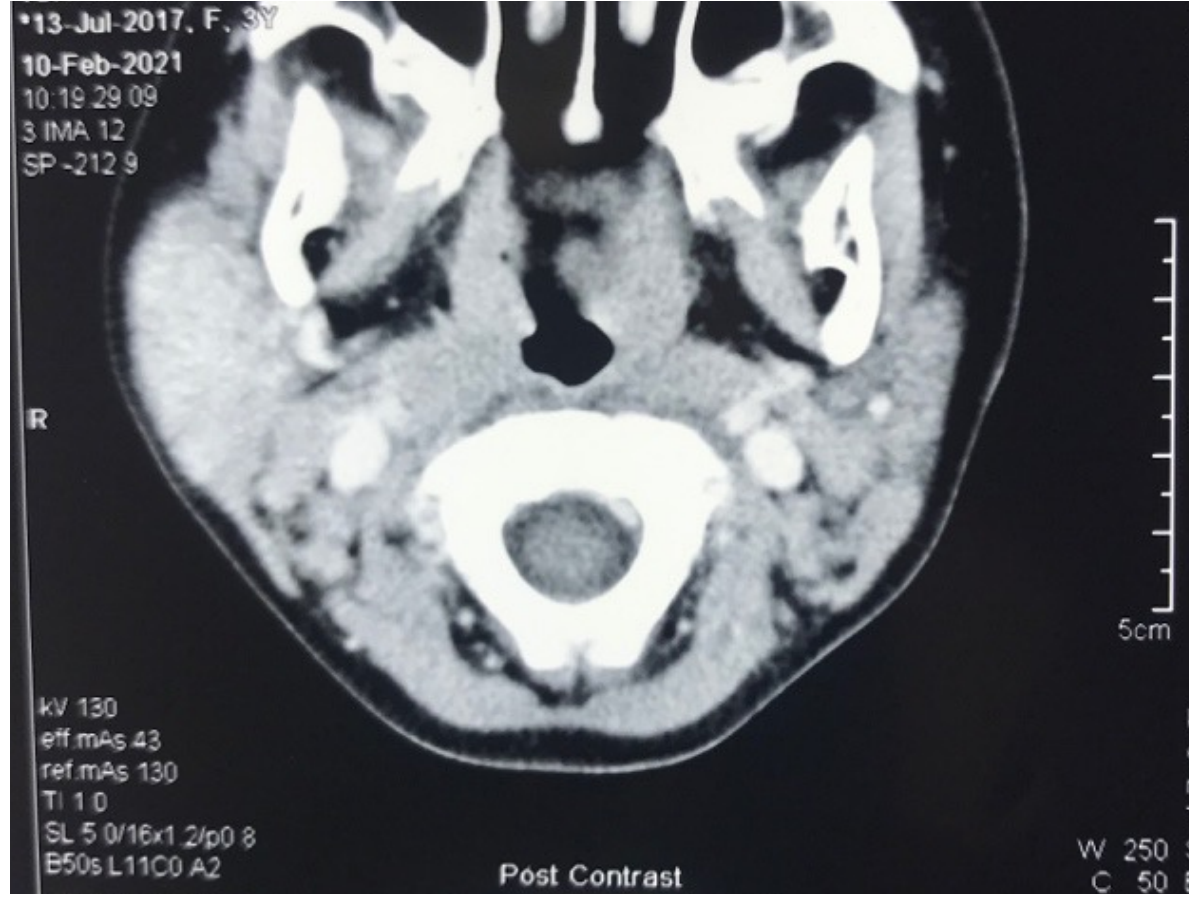


Fig 1: Axial CT of parotid mass

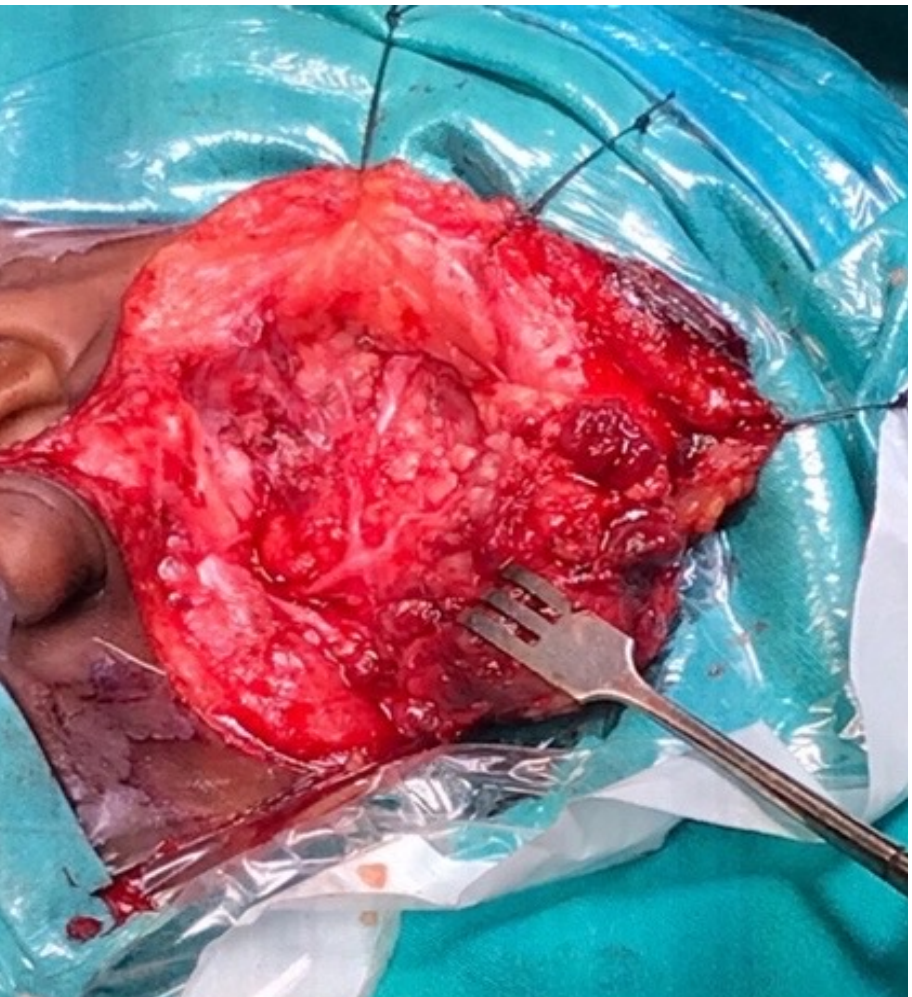


Fig 3: Superficial lobe of the parotid gland removed with exposure of the facial nerve and its branches

Discussion

- ❖ Salivary gland malignancies amount to less than 10% of all head & neck cancers in children.^[4]
- ❖ Incidence: low in all age groups at 0.4 to 2.6 cases per 100 000 in adults and 0.08 per 100 000 in children annually.^[2,5]
- ❖ Parotid masses are most likely to be malignant in children compared to adults ^[3] (50% and 15-32% in children & adults respectively) ^[3,6]
- ❖ MEC is the most common type of malignancy encountered in children.^[7,8]
- ❖ MEC is mostly low grade in children presenting at an average age of 13.5years with female predilection.^[3]
- ❖ Children under 10 years usually present with high grade MEC with poor prognosis.^[3,4]
- ❖ Our patient is a 3 year old female with an intermediate grade tumour which is atypical grading for her age group.^[3,4]

Aetiology

- ❖ Aetiology of MEC is unknown
- ❖ Genetics & prior exposure to ionizing radiation may play a role. ^[3,4]
- ❖ Other risk factors include:
 - Viral: Epstein Barr Virus(EBV), cytomegalovirus (CMV) infections
 - Environmental pollutants: asbestos, nickel, silica, rubber and woodworking material.^[4]
- ❖ Our patient was not directly exposed to any of these, though she lives in an area close to a platinum mine.

Clinical Presentation

- ❖ **History**
 - MEC presents as a painless mass with an average duration of 1-2 years.^[4]
 - Additional symptoms: pain, facial weakness, sleep disordered breathing, voice changes and hearing loss.^[2]
 - Our patient had a shorter duration of symptoms of 6 months with isolated episodic pain.

- ❖ **Examination**
 - Signs of malignancy e.g. facial nerve paralysis in up to 4% of cases. ^[2,9]
 - Tumor is typically firm, variable fixation, non-tender and size ranges from 1-3cm. ^[3]
 - Our patient did not exhibit any signs of malignancy. Facial nerve was intact & overlying skin were intact. The mass was non-tender.

Differential Diagnosis

- ❖ Congenital: Branchial cleft cyst, dermoid cyst
- ❖ Acquired:
 - Infective/Inflammatory: Acute parotitis, TB, lymphoepithelial cyst (in HIV)
 - Neoplastic:
 - Benign: Pleomorphic adenoma, haemangioma
 - Malignancies: MEC, acinic cell carcinoma, adenoid cystic carcinoma^[9,10]

Special Investigations

- ❖ Laboratory tests: full blood count, renal & liver function, serological testing (HIV, CMV and EBV).^[6]
- ❖ Imaging:
 - Radiographs: chest radiograph may reveal lung metastases
 - Ultrasound: distinguishes between cystic vs solid masses & helps for sonar guided FNAC biopsy. ^[2,9]
 - Cross-sectional imaging: comprehensive assessment of parotid masses, including involvement of the deep lobe or surrounding structures .^[6,9]
 - Magnetic resonance imaging (MRI): good for soft tissue characterization & delineating tumour margins ^[9], Facial nerve & perineural invasion better visualised with MRI. ^[6,9]
 - Computed Tomography (CT): good for identification of osseous involvement.^[9]
 - Positron emission tomography (PET) CT: Of more use to screen for metastatic disease.^[10]
- ❖ Tissue sampling:
 - FNA:
 - Invasive test for diagnosis of parotid tumours.
 - Although highly reliable may not be well tolerated by young children which can limit its use.
 - Accuracy of FNA to differentiate benign vs malignant masses is only 79%, & when malignancy confirmed unable to do immunohistochemistry & grading through cytology.^[2]
 - FNAC in our patient was inconclusive with diagnostic confirmation only on histopathological analysis post parotidectomy.
 - Biopsy:
 - Incisional/ excisional biopsy: Carries risks such as tumour seeding and facial nerve injury.^[2]
 - Core biopsy: For parotid tumors through planned incision in a safe region is an option in a few selected cases (suspected lymphoma, unequivocal FNA).^[2]

Histopathology

- ❖ Histopathological features of MEC for grading: nuclear atypia, cystic vs solid components, perineural +/- lymphovascular invasion, necrosis and mitotic activity.^[4,9]
- ❖ Grading systems:
 - Healy, Armed Forces Institute of Pathology and Brandwein (AFIP) stratify tumours into low, intermediate or high grade.^[9]
 - Low grade MEC: More cystic components
 - High grade MEC: Solid components
- ❖ Important in ultimate prognosis of paediatric MEC:
 - High grade MEC: poor prognosis & survival ^[9]
 - Low & intermediate grade MEC: good prognosis.^[9]
- ❖ The histology in our patient showed predominant solid growth pattern with 20% cystic component and was of an intermediate grade according to AFIP grading.

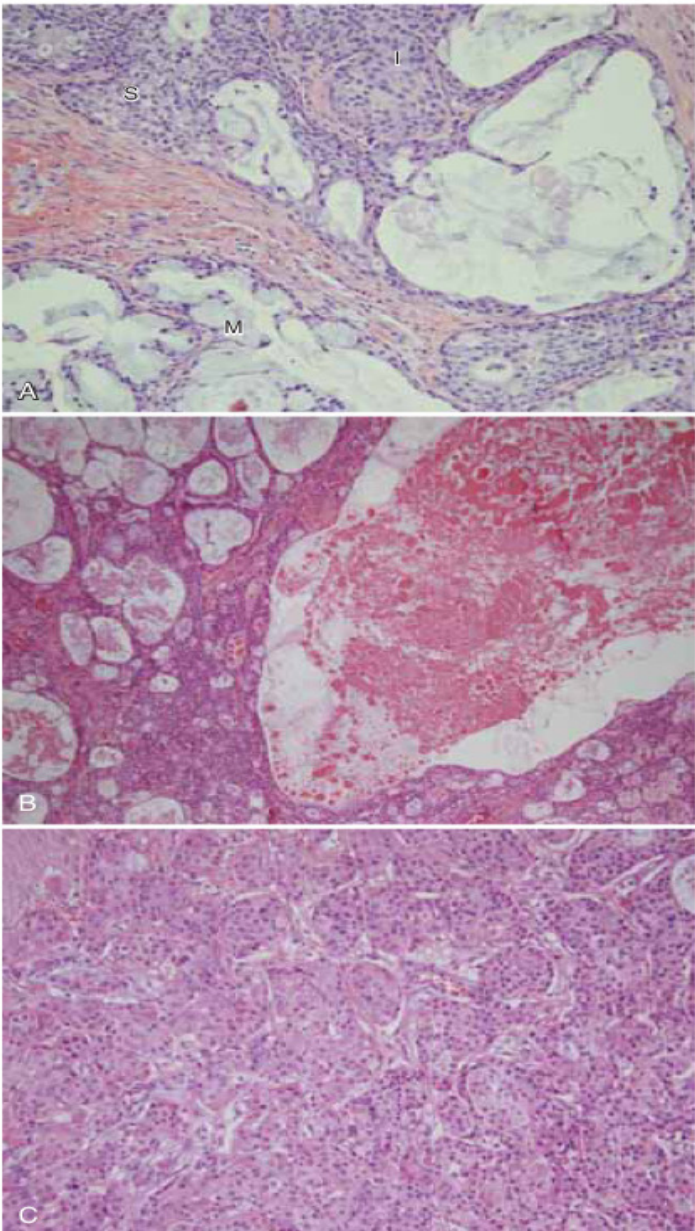


Fig 4: Diagram illustrating microscopy of MEC. Source: Cummings Otolaryngology Head & Neck Surgery^[7]

- A. Intermediate cells(I), intermixed mucous cells (M) and squamoid cells (S) or epidermoid cells with slightly more prominent pink cytoplasm.
- B. Tumor with extensive cystic change.
- C. High-grade MEC with solid sheets of intermediate cells, minimal mucous differentiation & no cystic change.

Staging

- ❖ Adult TNM staging is used in major salivary gland malignancies.^[5,9]
- ❖ Our patient was classified as stage III MEC (pT3N0M0) according to the TNM classification.

Management

- ❖ Surgery:
 - Parotidectomy:
 - Treatment of choice in paediatric MEC
 - Superficial or total depending on site of lesion with facial nerve preservation.^[2,4,7,8]
 - Good outcomes for children.^[4]
 - Indications for facial nerve resection:
 - ✓ Preoperative paralysis or intraoperative invasion by tumour.^[6]
 - ✓ Following resection: primary anastomoses or transpositional grafting indicated.^[9]
 - Neck dissection (ND):
 - Therapeutic ND: Indicated for clinical +/- radiological evidence of cervical metastases.^[4]
 - Our patient did not have any lymph nodes.
- ❖ Non-surgical:
 - Radiation:
 - In paediatrics given with caution in a select few due to potential adverse effects.^[2]
 - Intensity-modulated radiation preferred option due to sparing of normal tissues.^[8]
 - Indications: irresectable cases, positive margins, high-grade disease, perineural invasion, lymphovascular invasion & extraglandular extension.^[4]
 - Chemotherapy: Considered in recurrent disease or palliative cases.^[4,8]
 - Outcomes chemoradiation uncertain due to the rarity of these cancers in children & use is controversial.^[4]
- ❖ Our patient was managed surgically with a superficial parotidectomy with facial nerve preservation. Histopathological analysis revealed an intermediate grade MEC with clear margins with no indication for post-operative chemoradiation.
- ❖ No recurrence has been observed in our patient to date and long-term follow up will be continued.

Conclusion

Salivary gland malignancies are rare in children with an incidence of less than 5% mainly affecting children older than 10years. MEC is the most common malignant type usually affecting the parotid gland. The prognosis of MEC in children is usually good due to the low or intermediate grading commonly found in this age group. Primary surgery is usually sufficient & recurrence is rare.

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