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RESEARCH ARTICLE

EXTRAOCULAR SPREAD TO THE PAROTID FROM MEDULLOEPITHELIOMA IN AN 8-YEAR-OLD: CASE REPORT AND LITERATURE REVIEW

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Abstract

Introduction: Medulloepithelioma is a rare embryonal tumor that typic ally arises from the ciliary body in early childhood. Extraocular spread to the parotid region is exceptional and often represents intraparotid nodal metastasis from an ocular primary. Misleading benign imaging appearances can delay diagnosis.

Case Report: An 8-year-old girl with prior left ocular surgery for WHO stage II teratoid medulloepithelioma (evisceration with conformer, 2021) presented with a 1-year history of a progressive, painless left cheek swelling. Examination found a ~7 cm firm, fixed parotid mass with a ~1–1.5 cm ipsilateral upper jugulo-carotid node; no initial facial palsy.Magnetic Resonance Imaging (02/09/2024) showed a 66 mm lesion involving superficial and deep parotid lobes,favored as pleomorp hic adenoma.Core biopsy(30/10/2024)demonstrated medulloepitheliom a. Computed Tomography staging (07/11/2024) classified the disease T3N2bM0. She underwent total left parotidectomy with selective neck dissection(levels IIa IIb)on12/2024.Histology confirmed medulloepithe lioma with foci of necrosis, margin <1 mm, and 1/27 positive nodes without extracapsular extension or lymphovascular invasion. Multidisci plinary board recommended adjuvant radiotherapy, delivered to 60 Gy in 30 fractions(26/02/2025–11/04/2025).Post-Raditherapy CT(04/2025) showed no disease. Hematologic work-up:cytospin normal; marrow hypercellular/heterogeneous with 2% blasts; trephine hypoplastic; no extrahaematopoietic cells.Adjuvant chemotherapy(JOE regimen: carbo platin–etoposide–vincristine) was planned for six 21-day cycles (cycle 1 Day-1 on 08/05/2025). During cycle 5 she developed vomiting and a new peripheral facial palsy;she remained afebrile and hemodynamically stable; imaging was stable.

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Conclusion: In children with a parotid mass and a history of ocular disease, intraparotid metastasis from medulloepi thelioma should be considered. Tissue diagnosis is essential when imaging suggests benign salivary pathology.Multi modal therapy (surgery, adjuvant radiotherapy, and systemic therapy) provided early disease control in this case.

Introduction:-

Medulloepithelioma (ME) is an embryonal neuroepithelial tumor, usually originating from the non-pigmented ciliary epithelium and presenting in the first decade of life [1, 2]. Most eyes are confined-disease at diagnosis; extraocular spread worsens prognosis [3,4]. Parotid-region involvement is extraordinarily rare and typically reflects metastasis to intraparotid lymph nodes from an ocular primary rather than a true salivary-gland primary [5–7]. We describe an 8-year-old with parotid disease proven as ME, outline management, and contextualize findings with the literature.

Case Report:

An 8-year-old girl with normal development and vaccinations underwent left ocular evisceration with conformer placement on 16 September 2021 at an outside hospital. The outside pathology report favored teratoid medulloepithelioma (≥ WHO grade II).

Approximately one year before referral, a painless, progressively enlarging left cheek/parotid swelling developed without fever or constitutional symptoms. On examination she was WHO/ECOG 0. A ~7–8 cm firm, fixed left parotid mass was palpable, with a ~1–1.5 cm ipsilateral upper jugulo-carotid lymph node of similar consistency; there was no facial nerve palsy.

MRI (02 September 2024) showed a 66 mm lesion involving both superficial and deep lobes of the left parotid without clear local invasion; the pattern was initially read as compatible with pleomorphic adenoma. Parotid core biopsy (30 October 2024) established medulloepithelioma. Contrast-enhanced CT of chest/abdomen/pelvis (07 November 2024) demonstrated regional nodal disease with clinical–radiologic stage T3N2bM0 (salivary staging) (Figure 1&2).

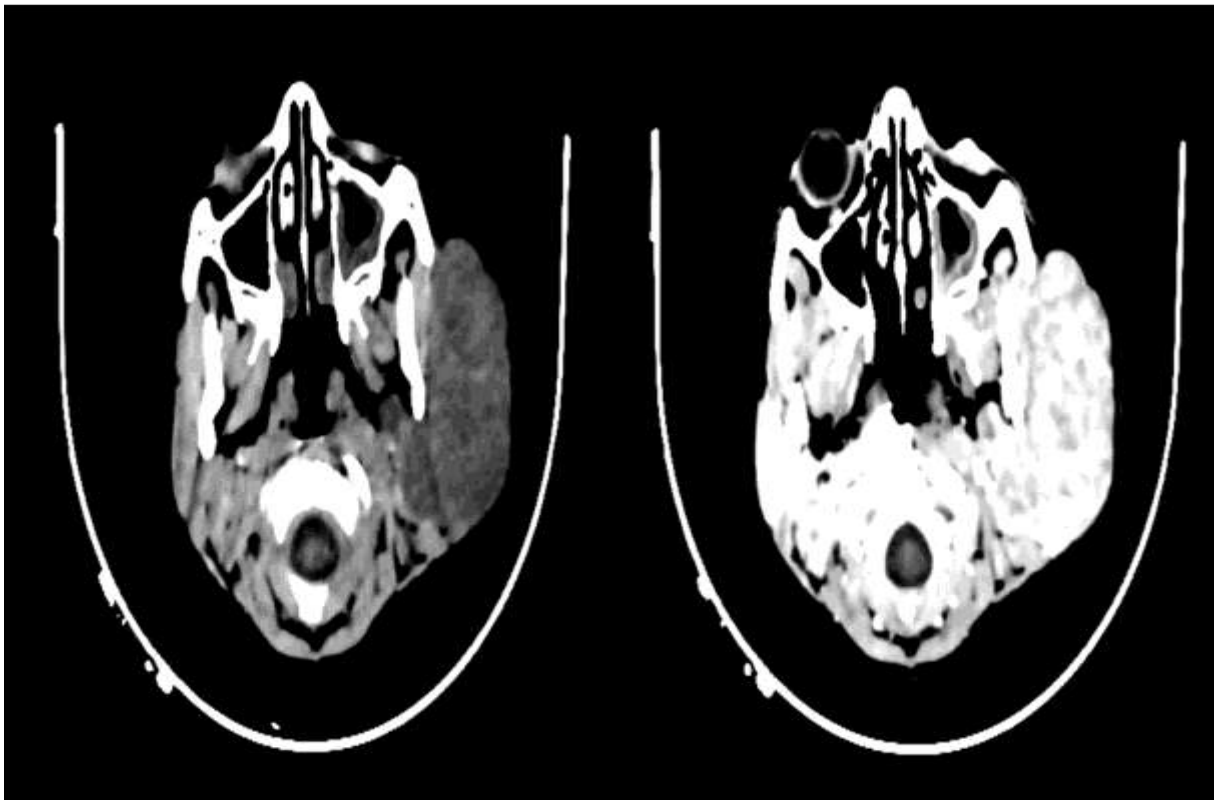

Figure 1: Axial CT images before and after contrast injection showing the mass involving the right parotid gland.

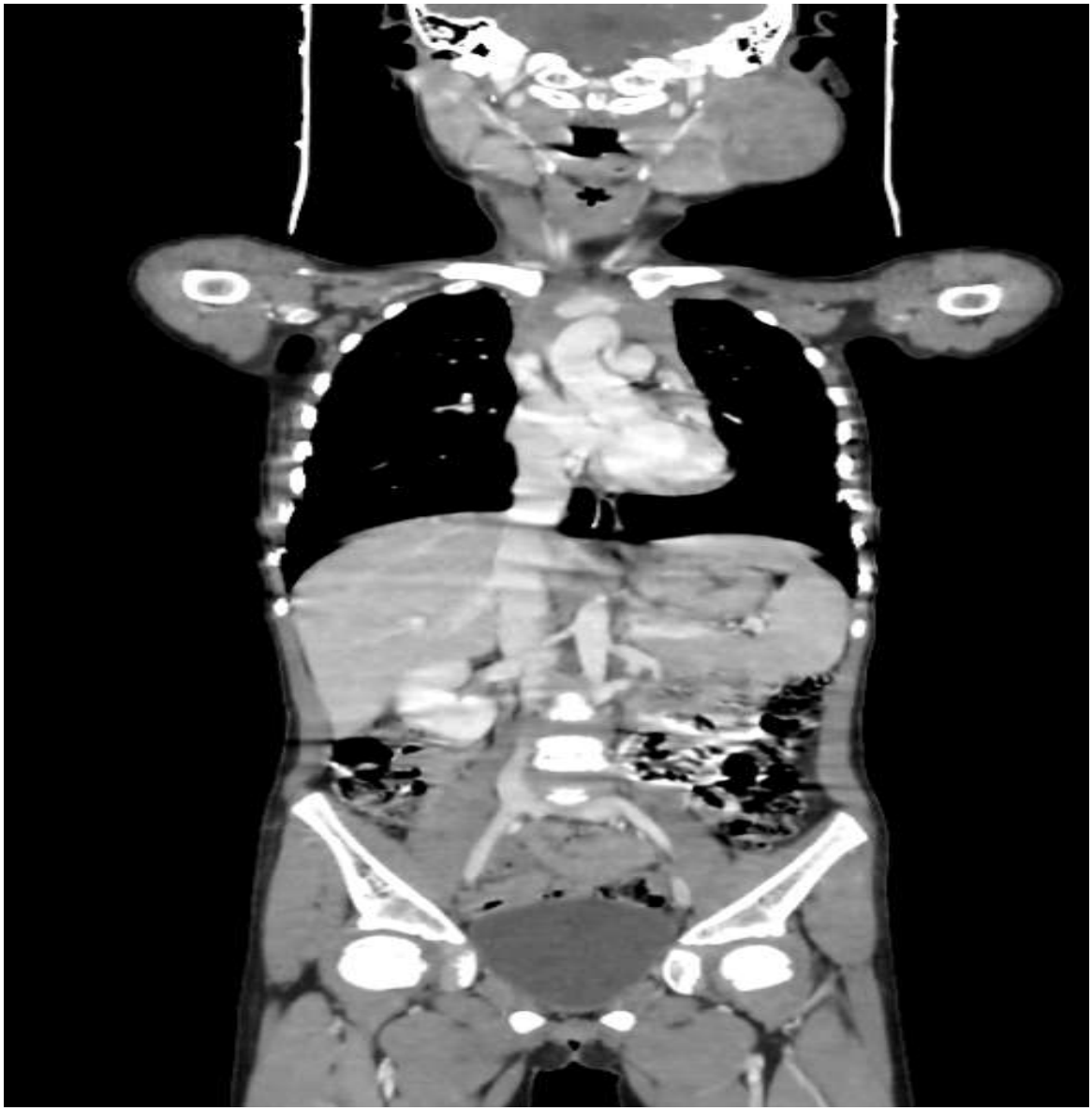

Figure 2 :Coronal view of a neck–chest–abdomen–pelvis CT showing the parotid tumor with no distant metastases.

On 23 December 2024, she underwent total left parotidectomy with selective neck dissection (levels IIa–IIb). Final pathology confirmed medulloepithelioma with necrosis, close surgical margin <1 mm, and 1 of 27 lymph nodes positive, without extracapsular extension or lymphovascular invasion.

A multidisciplinary tumor board (02 January 2025) recommended adjuvant radiotherapy to the parotidectomy bed and at-risk nodal levels. External-beam 3D radiotherapy was delivered to 60 Gy in 30 fractions from 26 February to 11 April 2025. Acute effects included radiodermatitis and localized radio-induced alopecia with secondary impetiginized dermatophytosis, treated medically. Post-RT CT (18 April 2025) showed no disease (Figure 3).

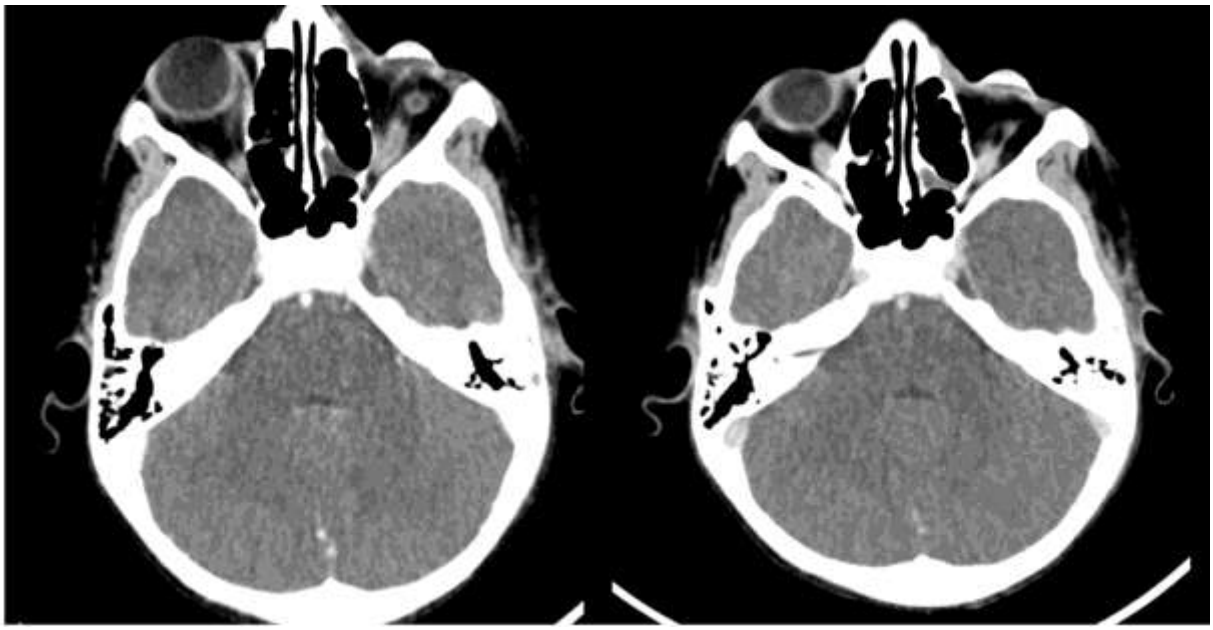

Figure 3: Axial CT images post radiotherapy showing complete clinical response.

Given the extraocular setting, adjuvant systemic therapy (JOE-type: carboplatin, etoposide, vincristine) was planned for six 21-day cycles, with cycle 1 day 1 on 08 May 2025. By mid-July 2025 she was receiving cycle 5. On 14 August 2025, she presented with vomiting and a new left peripheral facial palsy; she was afebrile, hemodynamically and respiratorily stable, and the parotidectomy scar was clean. CBC (11 August 2025) showed Hb 9.6 g/dL, WBC 4.1 × 10⁹/L (ANC 2.22 × 10⁹/L; lymphocytes 1.36 × 10⁹/L), platelets 214 × 10⁹/L. CT (14 July 2025) demonstrated no mass orabnormal enhancement in the left orbit and overall stable imaging. Cycle 6 was scheduled for 02 September 2025 with routine pre-cycle labs.

Discussion:-

Medulloepithelioma is an embryonal neuroepithelial tumor that most commonly arises from the non-pigmented ciliary epithelium of the eye in early childhood; extraocular spread is uncommon and portends a worse prognosis [1–4]. When the parotid region is involved, published experience indicates that the disease usually represents metastasis to intraparotid lymph nodes from an ocular primary rather than a true primary salivary-gland neoplasm [5–7]. This pattern is clinically important because a new pediatric parotid mass in a child with prior ocular pathology should prompt consideration of intraparotid nodal metastasis from medulloepithelioma.

This case also highlights a diagnostic pitfall: salivary imaging can suggest benign lesions such as pleomorphic adenoma, yet tissue diagnosis remains crucial in atypical contexts. In our patient, MRI favored pleomorphic adenoma, whereas core biopsy established medulloepithelioma, preventing delay in definitive management. Histologically, medulloepithelioma shows primitive neuroepithelial tubules and rosettes with variable neuronal or glial differentiation, and immunophenotyping often demonstrates vimentin and neuronal markers with variable cytokeratin expression, features that help separate it from salivary myoepithelial-lineage tumors [2].

Management of extraocular disease is multidisciplinary. Historical and contemporary reports support surgical control of intraparotid disease with parotidectomy and appropriate neck management, coupled with definitive treatment of the ocular primary when present; adjuvant radiotherapy and systemic therapy are considered based on margins, nodal status, and extent of spread [5–7]. In the present case, a close margin and nodal involvement justified adjuvant radiotherapy to 60 Gy, and systemic therapy was added given the extraocular setting. Short-term post-radiotherapy imaging showed no residual disease, and treatment-related events—including radiodermatitis and a later peripheral facial palsy during chemotherapy—were handled conservatively. Vigilant long-term follow-up is warranted to assess durability of control and late effects.

Conclusion:-

Parotid-region ME in children is exceedingly rare and typically metastatic from an ocular origin. This case emphasizes the need for biopsy of atypical pediatric parotid masses, and supports multidisciplinary, multimodal therapy for extraocular disease.

Conflict of Interest:
None declared.

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