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Pediatric Infratemporal Fossa Meningioma: Uncommon Presentation, Diagnostic Challenges, and Surgical Strategies

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Abstract **Case Report**

Meningiomas, common tumors of the central nervous system, rarely manifest in extracranial locations, and their occurrence in children is exceptionally rare. This paper presents a comprehensive case study of a 10-year-old female patient diagnosed with an infratemporal fossa meningioma. Immunohistochemistry played a pivotal role in confirming the diagnosis, while CT scans and MRI aided in assessing tumor extension and planning the surgical approach. The tumor was successfully excised using a trans-mandibular osteotomy approach. A thorough literature review highlighted the scarcity of extracranial meningioma cases, particularly in children, and revealed four hypotheses explaining their unusual extracranial localization. Surgical excision remains the primary treatment, with radiation therapy considered for partial removal cases. The diagnostic challenges posed by the paucity of symptoms and the pivotal role of histological confirmation were discussed. This case underscores the importance of immunohistochemistry for diagnosis and initiates the management process for extracranial meningiomas, shedding light on their atypical clinical presentations and etiopathology.

Keywords: Meningiomas, Extracranial Location, Infratemporal Fossa, Immunohistochemistry, Surgical Excision.

INTRODUCTION

Meningiomas, traditionally recognized as intracranial neoplasms arising from the arachnoid cap cells of the meninges, have long captured the attention of medical professionals and researchers due to their prevalence and intriguing biological behavior. However, in addition to their well-established intracranial presence, there exists a subset of these tumors that defy convention by manifesting in extracranial locations, presenting unique diagnostic and management challenges. These extracranial meningiomas, while relatively rare, contribute to the broader understanding of meningioma biology, clinical behavior, and treatment paradigms.

The emergence of meningiomas outside the confines of the cranium opens avenues of exploration into their pathogenesis, histological diversity, and clinical outcomes. The complexity of these tumors extends beyond their unique anatomical presentation, encompassing a spectrum of histological subtypes and variable clinical manifestations. These extracranial counterparts of meningiomas beckon a reevaluation of diagnostic approaches, therapeutic interventions, and prognostic considerations that may diverge from the well-established norms governing intracranial meningiomas.

The Extracranial meningioma of the infratemporal fossa is very rare, and this presentation is even more uncommon in a pediatric population. The challenge in this particular case is the diagnosis assessment and the surgical management because of its anatomical location. We report the case of 10-year-old female patient with an infratemporal fossa meningioma from the diagnosis assessment to the surgical management.

CASE REPORT

We present the case of a 10-year-old patient, identified as C.L., with no pathological medical history devoid of head or facial trauma, as well as radiation exposure. Three years prior to admission, the patient exhibited a progressively enlarging left jugal swelling accompanied by ipsilateral hypoacusis, exophthalmos, and migraines. Notably absent were facial palsy, tinnitus, diminished visual acuity, and neurological deficits (Figure 1).

Upon examination in July 2019, a firm, immobile, and painless left jugal mass extending to the temporal and parotid region, measuring 7 cm, was observed. The mass presented no signs of inflammation and was associated with grade I exophthalmos and asymmetrical soft palate findings. No facial palsy, palpable adenopathy, or neuromeningeal signs were noted. A normal ophthalmological examination was recorded. Comprehensive biological evaluation yielded no abnormalities.

Subsequent cervico-facial computed tomography (CT) in 2019 unveiled a lesion measuring 35*30*40 mm within the left infra-temporal fossa, extending cranially through the skull base, enhanced upon contrast medium injection (Figures 2-5). A facial MRI revealed a large, hyper-vascular infratemporal mass infiltrating facial and neck meninges, accompanied by temporal, left orbital, and endocardial extensions (Figures 6-7).

Histology and immunohistochemistry following mass biopsy yielded a diagnosis of grade I meningothelial meningioma according to WHO 2016 classification. An initial neurosurgical intervention achieved the excision of the entire endocranial portion of the tumor.

Postoperative MRI conducted in April 2021 disclosed the persistence of an irregular process centered in the left infra-temporal fossa. A significant meningeal implantation on the skull base measuring approximately 75*55*69 mm was evident in T1 iso-signal and T2 hyper-signal sequences, displaying heterogeneous enhancement following contrast medium injection. The tumor infiltrated deep facial and neck spaces, displacing the optic nerve and left eyeball (Figures 8, 9).

Surgical access to the infra-temporal fossa was achieved through a trans-mandibular lateral approach. Commencing with a temporal flap extended pre-auricularly along the lines of the initial incision, a superficial parotidectomy enhanced visualization of the facial nerve. Subsequent reflection of the temporalis muscle anteriorly and inferiorly maintained adequate blood supply. The master muscle dissection exposed the mandible. Transversal mandibular osteotomy and coronoidectomy enabled tumor exposure, revealing posterior retro parotid space infiltration, anterior orbital fat involvement with internal orbital wall bone lysis, posterior maxillary sinus wall engagement, and superior skull base contact. Total tumor excision was performed.

Mandible and coronoid process repositioning and fixation were accomplished using plates and screws. Layered wound closure ensued. Post-surgery, the patient presented with facial palsy of grade III in the House-Brackmann classification, receiving corticosteroids. Progression was marked by a regression to grade II (Figure 11).

Four weeks post-surgery, a facial CT scan indicated residual tumor persistence within the left infra-temporal fossa, with significant meningeal base implantation at the skull base and local infiltration of the left hemiface's deep spaces, including orbital extension (Figure 10).

A subsequent surgical revision, guided by neuronavigation, achieved subtotal tumor resection. Removal encompassed the tumor's meningeal base of implantation, the external orbital rim, and the residual tumor at the infra-temporal fossa level, extending to the large wing of the sphenoid. Neuronavigation provided procedural guidance, sparing the intraorbital tumor component (Figures 12-13).

Postoperative follow-up exhibited facial edema and a persistent grade II facial palsy. Discharge occurred four days post-surgery with oral antibiotics (amoxicillin-clavulanic acid) and corticosteroids. The patient's next course of action involves referral for complementary radiotherapy.

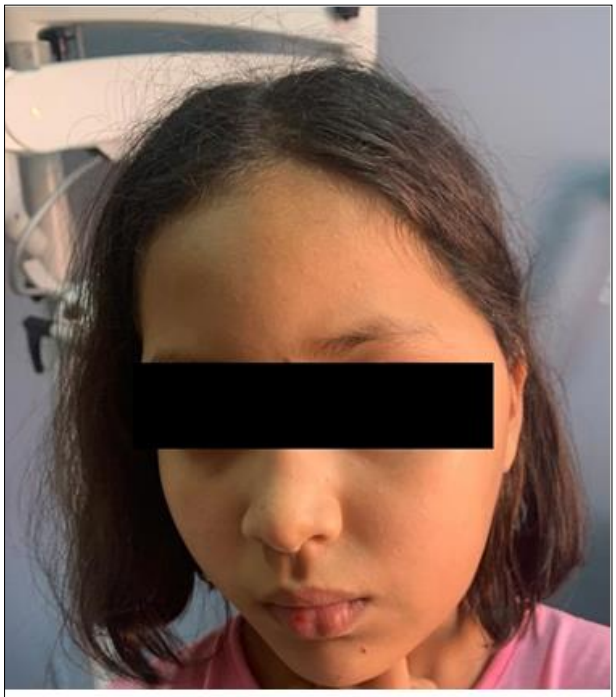


Figure 1: 10-year-old female patient with a left jugale swallowing.

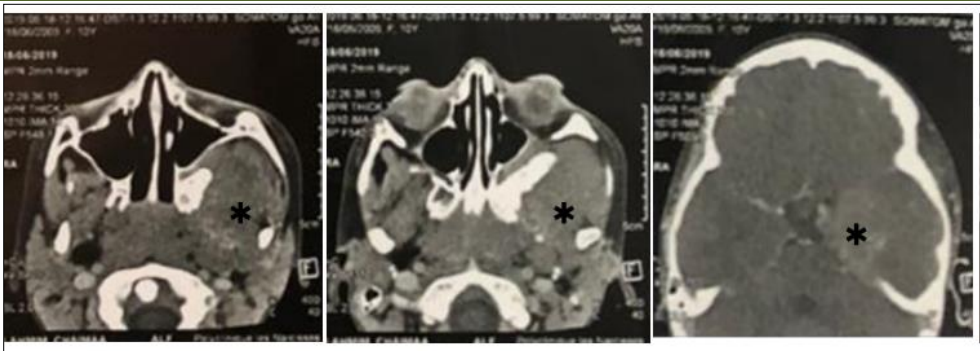


Figure 2.3.4 : axial section of the facial CT showing a lesion process centred on the left infra-temporal fossa with intra orbital and endocranial extension

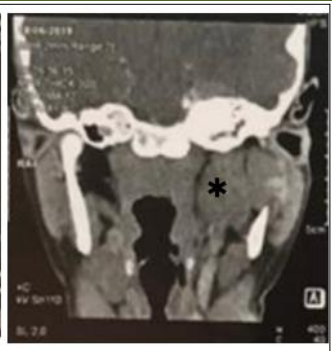


Figure 5: coronal section of the facial CT showing a lesion process centred on the left infra-temporal fossa.

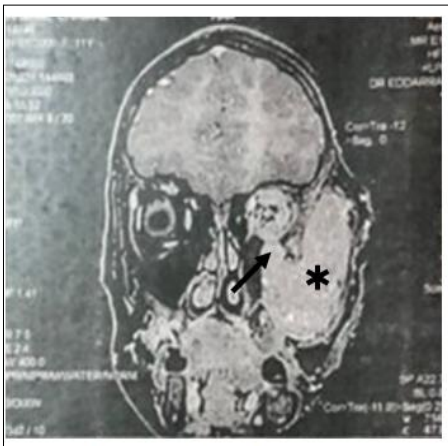


Figure 6: Facial MRI in frontal section showing a voluminous lesion process centred on the left infra-temporal fossa extended to the left orbital fat in T1 iso signal.

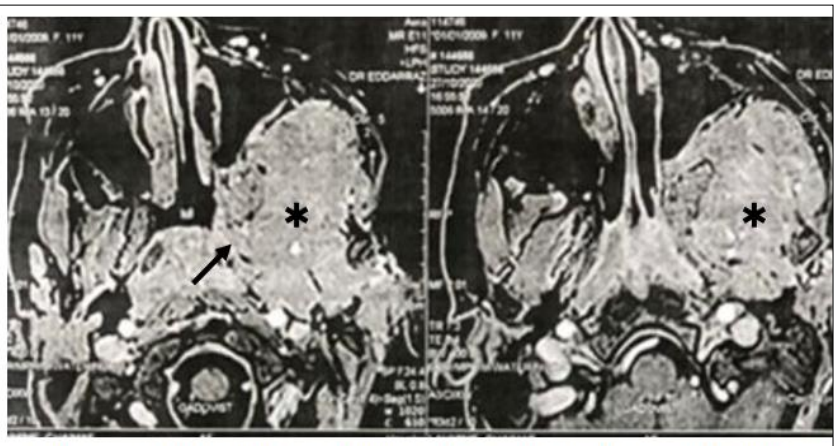


Figure 7: Facial MRI in axial section showing a voluminous lesion process centred on the left infra-temporal fossa in T1 Iso signal.

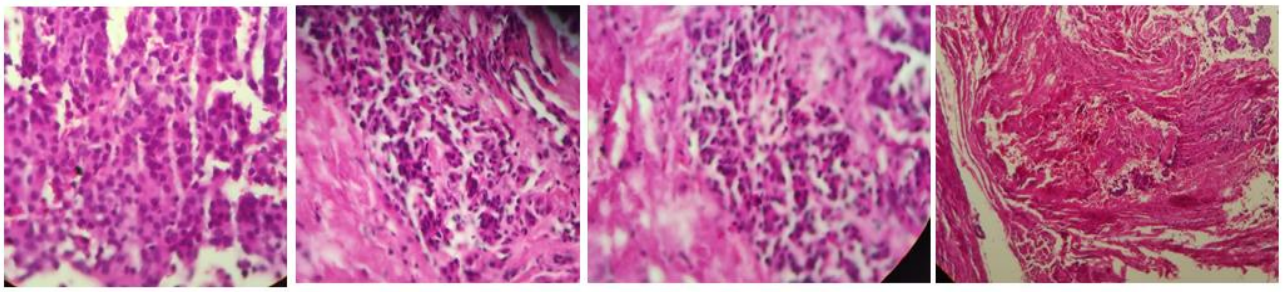


Figure 8: Microscopic pictures of the pathological study of the biopsy showing signs of meningothelial meningioma grade I according to the Who classification 2016 .

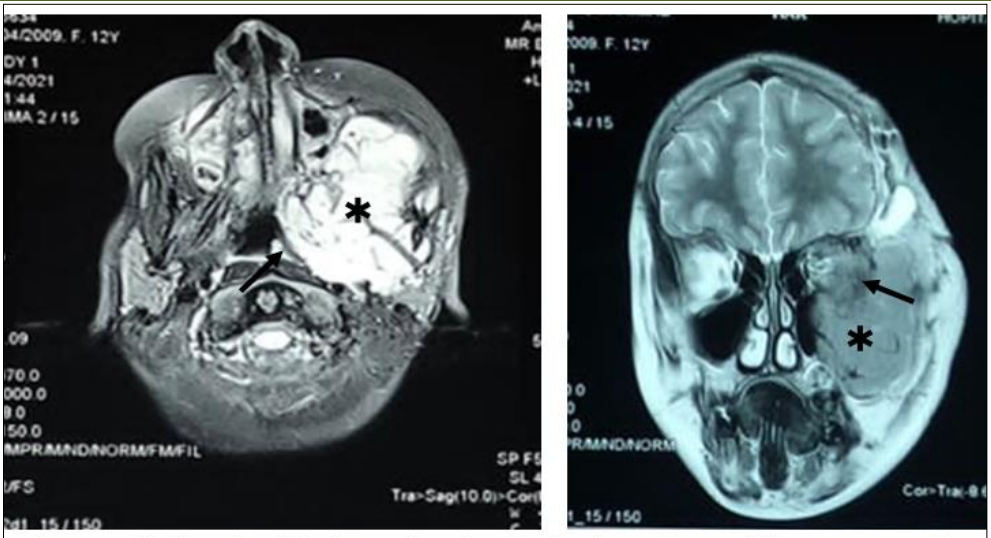
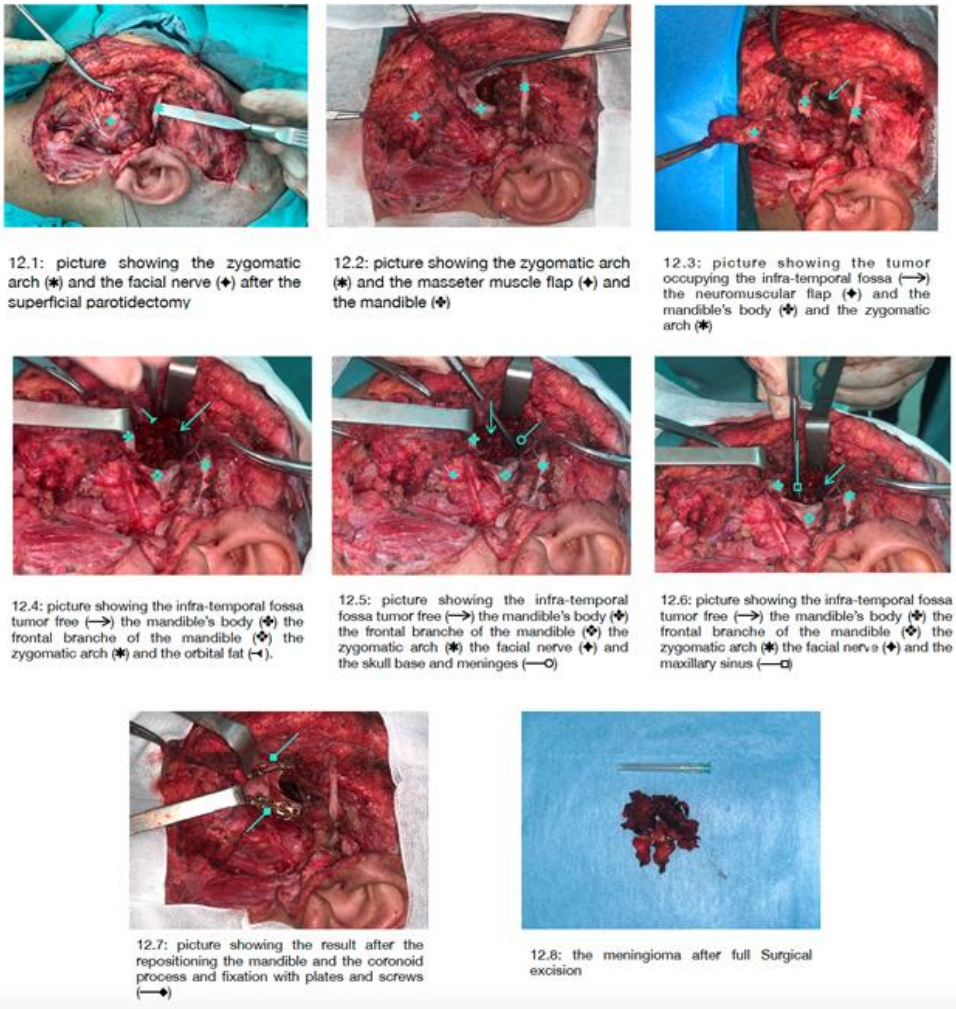


Figure 9.10: facial MRI in axial and coronal sections showing the persistence of the tumor process in the left infra-temporal fossa with large meningeal implantation and left intra orbital extension in T1 iso signal and T2 hyper-signal



12.1: picture showing the zygomatic arch (✱) and the facial nerve (✦) after the superficial parotidectomy

12.2: picture showing the zygomatic arch (✱) and the masseter muscle flap (✦) and the mandible (✤)

12.3: picture showing the tumor occupying the infra-temporal fossa (⟶) the neuromuscular flap (✦) and the mandible's body (✤) and the zygomatic arch (✱)

12.4: picture showing the infra-temporal fossa tumor free (⟶) the mandible's body (✤) the frontal branche of the mandible (✥) the zygomatic arch (✱) and the orbital fat (⊣).

12.5: picture showing the infra-temporal fossa tumor free (⟶) the mandible's body (✤) the frontal branche of the mandible (✥) the zygomatic arch (✱) the facial nerve (✦) and the skull base and meninges (⟵○)

12.6: picture showing the infra-temporal fossa tumor free (⟶) the mandible's body (✤) the frontal branche of the mandible (✥) the zygomatic arch (✱) the facial nerve (✦) and the maxillary sinus (⟵□)

12.7: picture showing the result after the repositioning the mandible and the coronoid process and fixation with plates and screws (⟵●)

12.8: the meningioma after full Surgical excision

Figure11: per-operative pictures of the superficial parotidectomy and transmandibular osteotomy to access the infra-temporal fossa and the tumor excision.

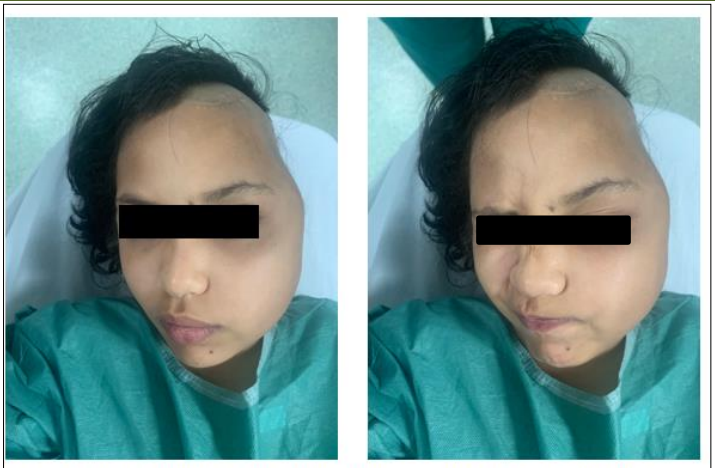


Figure 12: Picture of post operative facial palsy regression 1 month after the first surgery

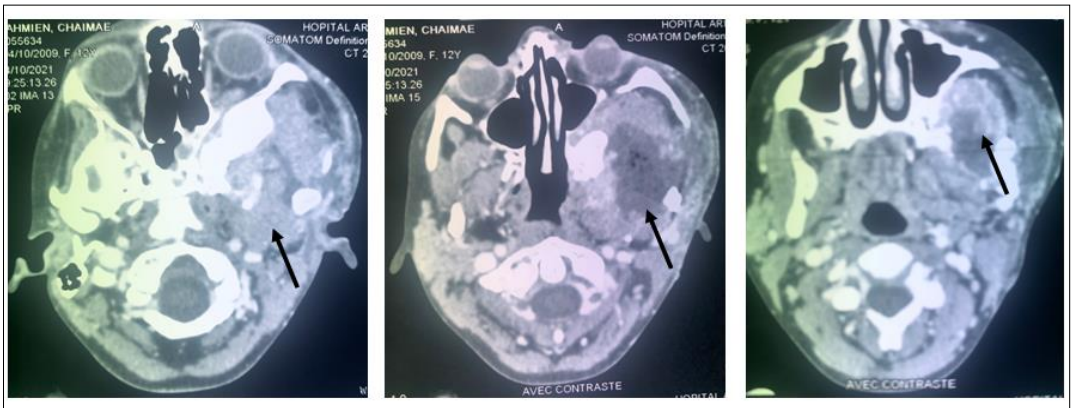
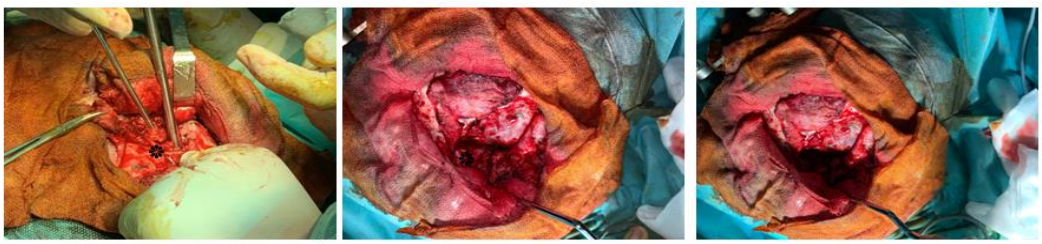


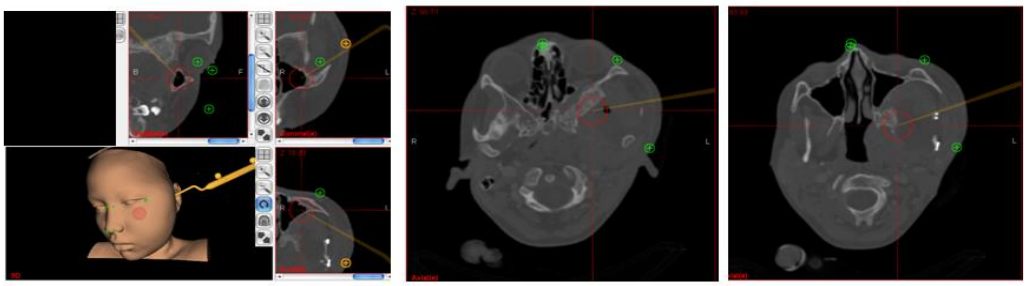
Figure 13: Post operative facial CT Scan showing a persistence of the tumour residue in the left infra-temporal fossa with a large meningeal base of implantation at the base of the skull, locally infiltrating the deep spaces of the left hemiface with orbital extension



per-operative picture showing the tumour (✿)

per-operative picture after a partial resection of the tumour (✿)

per-operative picture after a total resection of the tumour



neuronavigation control guided the resection throughout the procedure; The intraorbital part of the tumour was not resected

Figure 14: per-operative pictures of the second surgery using neuronavigation

DISCUSSION

Constitute the second most prevalent tumors of the central nervous system, accounting for 20-30% of all intracranial neoplasms [1]. These tumors predominantly affect female patients during their second decade of life [2]. In the pediatric population, meningiomas are less frequent than in adulthood, comprising only 2.2% of all childhood and adolescent neoplasms [3, 4]. The occurrence of extracranial meningiomas is a rarity, constituting merely 1-2% of all meningiomas [1-5].

The categorization of extracranial meningiomas was introduced by Hoye and colleagues in 1960 [6], distinguishing between primary and secondary forms. Primary extracranial meningiomas encompass two distinct types: those originating from arachnoid cells persisting within nerve sheaths, and those occurring ectopically without any connection to the cranial cavity. In contrast, secondary extracranial meningiomas are divided into two classifications: tumors that directly escape from the cranium where they originated, and extracranial meningiomas that have metastasized from intracranial sources [6, 7].

The cause of extracranial meningiomas is still not known, there are some possible hypothesis as sited by Rushing and al [2]:

1. Arachnoidal cells are present in the sheaths of nerves or vessels where they emerge through the skull foramina.
2. Displaced pacchionian bodies become detached, pinched off, or entrapped during embryologic development in an extracranial location.
3. A traumatic event or cerebral hypertension that displaces arachnoid islets.
4. An origin from undifferentiated or multipotential mesenchymal cells, such as fibroblasts, Schwann cells, or a combination of these, perhaps explaining the diverse pathologic spectrum found in meningiomas.

To conclude, the presence of arachnoid cells beyond the confines of the nervous system may instigate extracranial meningiomas [1-13]. Our case seems to align more fortuitously with the second hypothesis, as our patient, a 10-year-old child with an extracranial meningioma in the infratemporal fossa, lacks any history of cranial trauma, lending a particularly intriguing and atypical dimension to this case.

Diagnosing extracranial meningiomas poses challenges due to their paucisymptomatic and multifarious clinical presentation, often leading to misdiagnosis and delays in identification [2-9]. Notably, the cornerstone of diagnosis lies in histological confirmation, since interpretations of CT scans and MRIs may suggest alternative pathologies given the uncommon nature of extracranial meningiomas [1-14].

In our instance, the initial facial MRI scan suggested the potential of a rhabdomyosarcoma or an atypical nasopharyngeal fibroma, given the tumor's traversal of the sphenopalatine foramen. Subsequent histological and immunohistochemistry analyses definitively established the diagnosis as a meningioma originating from the infratemporal fossa.

Literature predominantly advocates surgical resection as the primary treatment, aiming for complete excision to mitigate recurrence risk. In instances of incomplete excision, adjunctive radiation therapy is recommended [4-15]. Our surgical approach, selected due to its extensive usage, granted comprehensive access to the infratemporal fossa, facilitating complete tumor excision, coupled with osteosynthesis [1-19].

Postoperative outcomes and prognosis are promising with full surgical extirpation, showcasing survival rates of up to 78% at 10 years [1-20]. Notably, Apra and colleagues [21], underscore that grade I meningiomas can be managed through exclusive surgery or radiosurgery. The consideration of adjuvant radiotherapy or radiosurgery arises for patients harboring growing remnants, a circumstance akin to that of our patient.

CONCLUSION

In conclusion, the case of an extracranial meningioma in a 10-year-old patient underscores the intricacies and challenges associated with this rare condition. The atypical presentation of such tumors outside the central nervous system, particularly in pediatric populations, accentuates the significance of accurate diagnosis and tailored management.

Diagnosing extracranial meningiomas proves intricate due to their subtle symptoms and the diversity of potential differential diagnoses. Histological confirmation emerges as a pivotal diagnostic tool, as radiological interpretations may not distinctly indicate these uncommon tumors.

Surgical resection stands as the primary therapeutic approach, emphasizing complete excision to minimize recurrence risks. When excision is partial, adjunctive radiation therapy plays a crucial role in achieving comprehensive treatment outcomes.

This case not only contributes to the limited literature on pediatric extracranial meningiomas but also underscores the importance of multidisciplinary collaboration, precision in diagnosis, and tailored therapeutic strategies. As knowledge and techniques evolve, further insights into the etiology, diagnosis, and treatment of extracranial meningiomas hold the promise of enhancing patient outcomes and refining clinical management.

Informed Consents

Informed consent was obtained from the patient prior to participation in the study. The patient was informed of the study's objectives, procedures, potential risks and benefits, and the right to withdraw at any time without consequence. All patient information was kept confidential and secure to protect their privacy.

Conflict of Interest: The authors declare no conflict of interest.

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