The Journal of Medical Research

***Case Report***

JMR 2023; 9(2):20-22
March- April
ISSN:2395-7565

www.medicinearticle.com
Received:12-03-2023
Accepted:26-04-2023
DOI: 10.31254/jmr.2023.9203

# A rare Case of Rhabdomyosarcoma presenting as Gradenigo syndrome in a child

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## Abstract

Objective: To study a rare case of Rhabdomyosarcoma. Methods: Observational descriptive study of a case, a 3-year-old female child, presenting to our OPD. She was thoroughly examined and radiological investigations (HRCT Temporal bone and MRI Brain) were done. The Neurosurgery department proceeded with a biopsy of the lesion by left temporal bone craniotomy through subtemporal approach. Results: Histopathology suggested a Rhabdomyosarcoma of anaplastic type. The patient then underwent 6 cycles of Chemoradiation. Conclusion: The awareness that Rhabdomyosarcoma of petrous apex can present as Gradenigo syndrome along with 7th nerve palsy is important for the early diagnosis and its treatment.

**Keywords:** Rhabdomyosarcoma, Gradenigo Syndrome, Petrous apex lesion.

## INTRODUCTION

Rhabdomyosarcoma is the most common soft tissue sarcoma in children [1]. It involves the external auditory canal and middle ear cavity and is also seen as an exophytic growth in the facial region. But in our case, there was no such evident mass seen thus making our case a rare presentation.

The petrous apex is a pyramid-shaped complex anatomic region formed by the medial portion of the temporal bone comprising a number of critical neural and vascular structures [2]. It is bounded laterally by the inner ear structures, the petro-occipital fissure medially, the petro-sphenoidal fissure and ICA anteriorly, and posterior canal fossa behind. The Dorello's canal extends through the posteromedial portion of the petrous apex and contains the abducens nerve.

The anterosuperior aspect of the petrous apex has a smooth depression that serves as the floor of the Meckel's cave which contains the Trigeminal or Gasserian ganglion and the rootlets of Trigeminal nerve.

Patients with petrous apex lesions present symptoms such as headache, retro-orbital pain or craniopathies due to involvement of IAC (8$^{th}$ nerve) like sensory neural hearing loss, tinnitus and vertigo, Diplopia (3$^{rd}$, 4$^{th}$ and 6$^{th}$ CN) and facial palsy (7$^{th}$ CN). The Trigeminal and Abducens nerves are susceptible to compression due to a thin layer of dura mater.

Petrous apex lesions can be classified on the basis of their origin into the following categories: developmental lesions, vascular lesions, inflammatory lesions, benign tumours, malignant lesions, and osseous dysplasia.

## CASE REPORT

A 3-year-old female child presented to our hospital with facial asymmetry for 1 month. She had developed a head tilt to the right side for the past 3 weeks. There was a history of headaches, retro-orbital pain and vomiting for the past 1 month. Also, there was one episode of giddiness.

History of decreased hearing or tinnitus could not be elicited.

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No history of any post aural swelling or earache.

**Fig 1:** Clinical picture of patient showing left side facial asymmetry and left lateral rectus palsy

On examination, there was facial asymmetry on crying, incomplete closure of left eye, loss of wrinkling on the left side of her forehead, deviation of angle of mouth right side and left grade 4 facial palsy LMN type with a left lateral rectus palsy. The left eye was deviated towards the medial canthus on central gaze. On otoscopic examination, the pre and postauricular regions the left side were normal. The external auditory canal was filled with discharge and after suctioning, the tympanic membrane was visualized. There was a grade 2 pars tensa retraction with an intact tympanic membrane. The right ear examination was normal with an intact tympanic membrane.

The left eye had mucopurulent discharge, ptosis, incomplete closure of eye, bell's phenomenon, conjunctival congestion, exposure keratitis of the inferior cornea, absence of corneal reflex. The loss of corneal reflex suggested an impairment of the afferent pathway in the ophthalmic division of the Trigeminal nerve. Paraesthesia over left maxillary region was also present. Gag reflex was present. No deviation of tip of the tongue. No change in voice. There were no signs of meningeal irritation. Hence, our initial clinical diagnosis was left unsafe CSOM with left side III, IV, VI and VII cranial nerve palsies caused by left petrous apicitis with cavernous sinus involvement.

**INVESTIGATION**

Blood investigations on admission showed Hemoglobin 13.1, and leukocytosis 14.8 x10 /L of which 83 % neutrophils,16 % lymphocytes, and 1 % were eosinophils and platelet count 4 lakh . Her liver and renal function tests were normal.

The HRCT temporal bone showed a well-defined soft tissue density mass lesion in the left petrous apex, extending superiorly along the left inferior extradural space and inferiorly into left parapharyngeal and carotid space, medially causing erosion of the clivus, left lateral border of the body of sphenoid and jugular tubercle. Laterally there was erosion of the medial half of the left IAC and the medial wall of mandibular fossa. There is erosion of the anterior inferior wall of the tympanic cavity with opacification of the tympanic cavity.

The Carotid canal and jugular bulb are not seen separately. Pus culture sensitivity showed pseudomonas and staphylococcal infection. Therefore, she was started on iv antibiotics such as Amikacin, Metronidazole and Vancomycin.

On no improvement in symptoms, an MRI brain was done which was reported as T2 hyperintense and T1 Hypointense enhancing lytic expansile lesion measuring 2.9x 2.0 x3.1 cm epicenter in the petrous apex probably a high-grade sarcoma or round cell tumour.

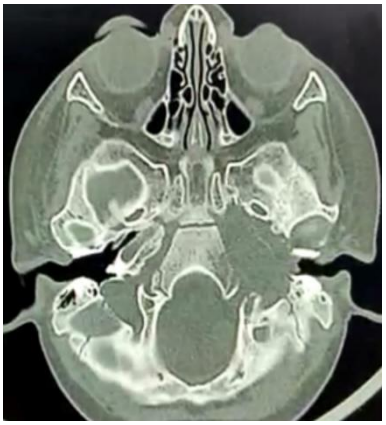

**Fig 2:** HRCT Temporal bone showing a left petrous apex soft tissue density

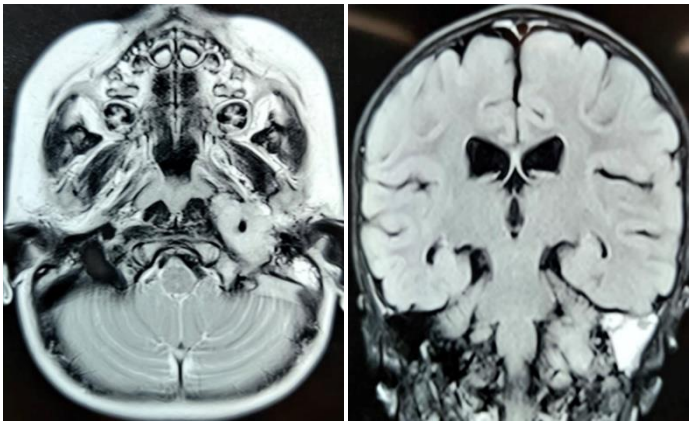

**Fig 3:** MRI scan showing Left petrous apex mass

Pt was referred to neurosurgery for a biopsy of the lesion. She underwent a left temporal craniotomy with biopsy through subtemporal approach. The histopathology was suggestive of primary embryonal rhabdomyosarcoma of anaplastic type. She is currently on regular follow-up and undergoing chemoradiotherapy

**DISCUSSION**

Rhabdomyosarcoma accounts for 7.8 to 25.7 % of cases among all head and neck cancers in children [1]. This tumour of the middle ear is a type of parameningeal rhabdomyosarcoma. The middle ear cleft is an unfavourable site as it is prone to many cranial nerve palsies, skull base erosion and intracranial extensions here. They present with complaints of otorrhea, hearing loss and aural polyp.

In our case, facial palsy was the main presenting feature along with intermittent ear discharge, there was no obvious polyp. Extensive neoplastic invasion of the skull base and cavernous sinus can result in multiple craniopathies causing complete ophthalmoplegia (3$^{rd}$, 4$^{th}$, 5$^{th}$ nerve palsy ]1]. In our case, there was lateral rectus palsy due to involvement of 6$^{th}$ cranial Nerve and it was evident by the deviation of left eye towards medial canthus and the head tilt to right.

Neuroimaging using CT and MRI are complementary modalities needed for accurate diagnosis. In our case, both were done and MRI was suggestive of an T2 hyperintense and T1 Hypointense enhancing lytic expansile lesion measuring 2.9x 2.0 x3.1 cm epicenter in the petrous apex probably a high-grade sarcoma or round cell tumour.

Gradenigo's syndrome is characterized by the triad of suppurative otitis media, deep seated retroorbital pain in the distribution of trigeminal nerve and lateral rectus palsy due to involvement of Abducens nerve [3]. It occurs secondary to apical petrositis which may be due to suppurative otitis media or extradural mass, or lesion in petrous apex.

The spread of inflammation may be via pneumatised air cells, vascular channels or directly through fascial planes.

The abducens nerve is particularly vulnerable in Dorello's canal, an inflexible channel roofed by Grubber's ligament. The trigeminal nerve is involved when the Gasserian ganglion gets affected in the Meckel's cave of the petrous apex. The management of Gradenigo syndrome is usually surgical and when not treated can result in meningitis, intracranial abscess, Vernet's syndrome, cavernous sinus thrombosis or hydrocephalus.

**CONCLUSION**

Awareness of the anatomy of the petrous apex and the varied presentation of pathology is useful in order to investigate and treat promptly. The presence of 7th and 8th cranial nerve palsy simultaneously should therefore raise the suspicion of petrous apex involvement. HRCT and MRI play an important role in its diagnosis and management is tailored depending on the suspected etiology.

**Contributions**: All the authors contributed in the preparation of this research and article.

**Conflicts of interest**

None declared.

**Financial support**

None declared.

**REFERENCES**

1. Muranjan M, Karande S, Parikh S, Sankhe S. A mistaken identity: rhabdomyosarcoma of the middle ear cleft misdiagnosed as chronic suppurative otitis media with temporal lobe abscess. BMJ Case Rep. 2014 Sep 19;2014:bcr2014206615. doi: 10.1136/bcr-2014-206615. PMID: 25240007; PMCID: PMC4170300.
2. Ahmed Abdel Razek, Benjamin Y. Huang, Lesions of the Petrous Apex: Classification and Findings at CT and MR Imaging, RadioGraphicsVol. 32, No. 1, https://doi.org/10.1148/rg.321105758
3. Thomas E Rossor, Yvonne C Anderson, Nicholas B Steventon, Lesley M Voss, Conservative management of Gradenigo's syndrome in a child, BMJ Case Reports 2011; doi:10.1136/bcr.03.2011.3978