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Peripheral Facial Palsy and Orbital Apex Syndroma as Complications of an Advanced Nasopharyngeal Paediatric Rhabdomyosarcoma

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ABSTRACT

Rhabdomyosarcoma is the most common soft tissue sarcoma encounered in childhood and adolescence; that arises from embryonic mesenchyme. It is most common found in the head and neck region, and the parameningeal location has the least favorable prognosis because of its tendency to local and intracranial extension. Parameningeal Rhabdomyosarcoma represents a diagnostic and therapeutic problem. We present the case of a child who got treated for her local nasopharyngeal rhabdomyosarcoma for over 1 month, and then got admitted for cranial nerve paralysis (peripheral facial palsy and Orbital apex syndrome) after the cranial extension of the tumor.

Introduction

Rhabdomyosarcoma (RMS) is an aggressive malignant tumor that arises from embryonic mesenchyme. It is the most common soft tissue tumour in children, with the head and neck region accounting for 35-40% of cases. It belongs to one of the most rapidly proliferating tumors and as it doesn't have a capsule, it may easily infiltrate surrounding tissues and distant organs by forming so-called skip lesions [1].

The prognosis of parameningeal Rhabdomyosarcoma (PM RMS) is still unsatisfactory, necessitating a more aggressive therapy, mostly due to delay in diagnosis, the tumor tendency to intracranial spread and to increased local recurrence. Its Treatment is complex, including multi-drug chemotherapy, radiotherapy and surgery.

We present the case of 9 years old child who's been treated for a rhabdomyosarcoma for over 1 month with chemotherapy, however it was complicated by a peripheral facial palsy and orbital apex syndrome "OAS".

Case report

We present the case of a 9 years old patient with no medical history, who was brought to our hospital for purulent rhinorrhea and chronic nasal obstruction for over 4 months associated with asthenia and weight loss. Nasopharyngeal endoscopy with biopsy showed a mass involving the entire nasopharynx. Histologic analysis with immunohistochimy revealed an embryonic rhabdomyosarcoma with a diffuse proliferation made of round cells with a high nuclear cytoplasmic ratio and hyperchromatic nuclei (Figure 1), and the tumor cell expressed desmin and myogenin (Figure 2). The child was referred to the paediatric oncologist and was prescribed chemotherapy. She received 2 cycles of IVA (ifosfamide, Vincristine, Adriamycin) and get discharged.

One month later, the patient presented a facial asymmetry with ptosis and got admitted in our ENT division. Clinical examination showed left eye ptosis with a right peripheral facial palsy grade III (House Brackmann score). Her ophthalmologic examination exhibited an orbital apex syndroma (OAS): low visual acuity in the right eye, accompanied by a complete right eye ophthalmoplegia, with no compromise in her colour vision (Figure 3).

Magnetic resonance imaging showed an organic process of the nasopharynx invading the infratemporal fossa, the pterygoide processes with the prevertebral,retrostyloid and vascular spaces, the middle ear and mastoid cells. The MRI showed an intracranial and Cerebellopontine Angle extension with an invasion of the trigeminal nerve too (Figure 4, 5).

The child was referred to the paediatric oncologist and was prescribed chemotherapy. It is well known that RM is chemosensitive, with the use of multiple drugs normally required. However, the patient didn't have a good evolution and then died 3 months later.

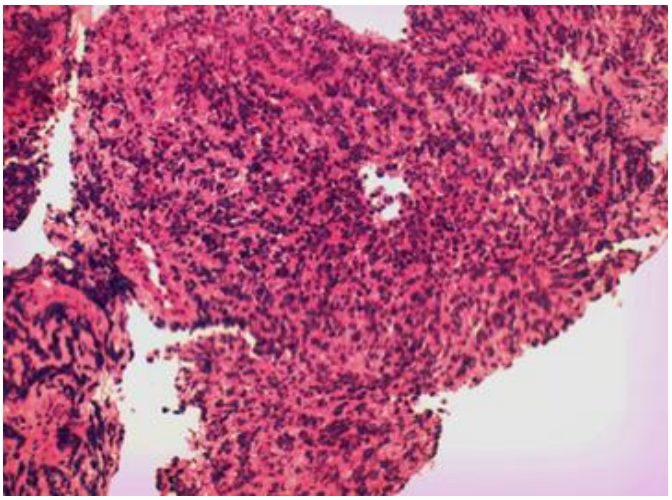

Figure1. Anatomopathologic examination revealed a diffuse proliferation made of round cells, with a high nuclear cytoplasmic ratio and hyperchromatic nuclei. HE 100x.

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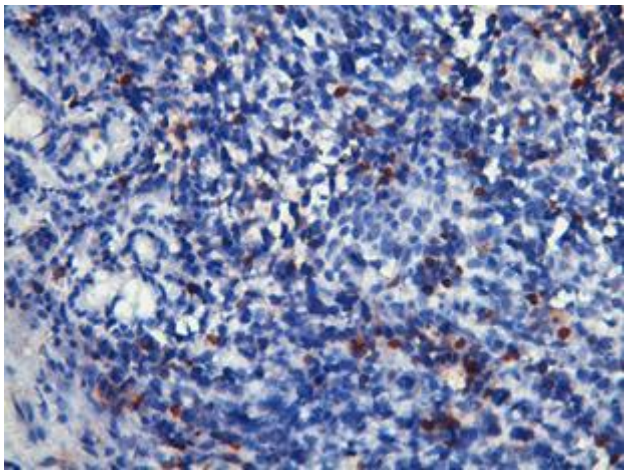

Figure2. The tumor cells express myogenin.

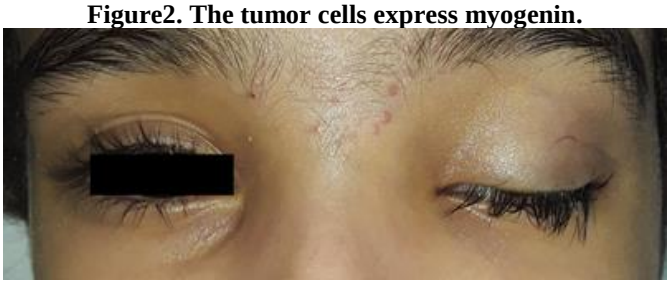

Figure3.Picture demonstrating the patient with left ptosis.

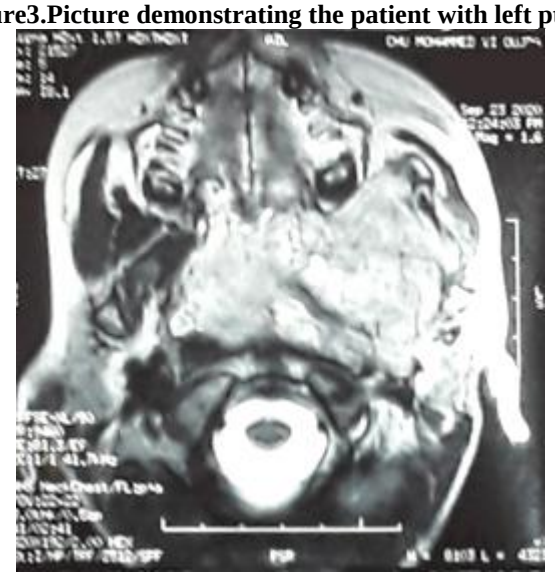

Figure4. Contrast enhanced T2 axial MRI image showing the nasopharyngeal mass involving the infratemporal fossa and the oropharynx.

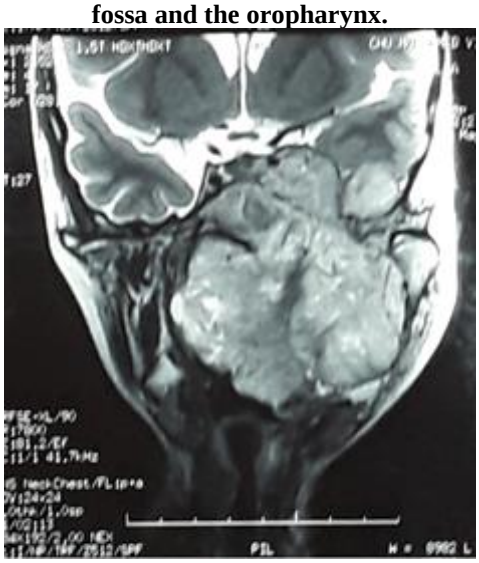

Figure5. Coronal MRI shows the intracranial extension of the process.

Discussion

Rhabdomyosarcoma is an aggressive malignant soft tissue tumor that arises from primitive striated muscle cells called rhabdomyoblasts. It is the most common soft tissue malignant tumor in children and accounts for about 7–8% of childhood cancers. It can arise from almost anywhere in the body. Most RMS in the pediatric population (30%-40%) occurs in the head and neck region [2].

Localization of the primary tumor has a prognostic value and therefore it determines the therapeutic management strategy. Three tumor sites can be distinguished in the head and neck region: parameningeal, orbital and the third one – meaning the rest of structures in the head and neck region. 25% of RMS are localized in the parameningeal region .This group includes tumors of the nasal cavity, nasopharynx, paranasal sinuses, middle ear and skull base. Parameningeal location has the least favorable prognosis because of the complexity of its anatomy, the proximity to the cranial cavity, the risk of dissemination via the cerebrospinal fluid, and also to the paucity of distinctive symptoms [3, 4].

Symptoms often mimic chronic upper respiratory tract infection, otitis media, and soft tissue injury such as nasal discharge, congestion, otorrhea, and mild swelling that may lead to the risk of misdiagnosis. The Late diagnosis is directly associated with poor prognosis [2, 4, 5].

Cranial nerve disorders or other neurologic symptoms that suggest skull base or central nervous system infiltration are an indication for urgent radiologic imaging.

Our patient had developed the OAS which characterized by lesions located at the posterior foramina of the bony orbit involving the superior orbital fissure and optic canal. The most common signs seen in the OAS are: Third, fourth, and sixth nerve pareses, sensory loss in the V1 distribution, and optic neuropathy from second cranial nerve involvement. The visual loss and ophthalmoplegia may precede any signs of orbital inflammation. Pain and proptosis can be associated too [6]. It can be explained in our case by the invasion of the adjacent structures (the cribriform plate, the brain and Cerebellopontine Angle), in the same time he developed a peripheral facial palsy because of the invasion of the middle ear, the filling of mastoid cells and Cerebellopontine angle.

The diagnosis of RMS should be based on the physical examination, past medical history, radiological imaging, histology, laboratory and molecular tests. Physical examination includes visualization and palpation of the face, scalp, oral cavity and pharyngeal structures, anterior and posterior rhinoscopy, microscopic ear examination, nasopharyngeal, hypopharyngeal and laryngeal endoscopy, evaluation of cranial nerve functions, eye and lymph node examination.

Magnetic resonance imaging (MRI) with contrast is the basic radiological imaging in the diagnosis of RMS of the head and/or neck. It can localize and measure precisely tumor size, evaluate local invasiveness, and visualize metastases to the lymph nodes as well as meninges and brain tissue infiltration. It also allows the evaluation of residual tumor mass after surgery and in diagnosis of tumor recurrence. CT scan with a contrast can evaluate the bone infiltration in the facial cranium and neurocranium [2].

Histologically, RMS are small round blue cell tumors – malignant low-grade tumors, typical for children. These tumors are composed of small cells, with a large, round, hyperchromatic nucleus that stains to dark blue under the influence of hematoxylin and eosin. Three basic types can be distinguished and their differentiation determines the therapy:

embryonal (ERMS) – the most frequent form (the same form in our case), alveolar RMS, and pleomorphic RMS especially in adult [7]

Rhabdomyosarcoma proliferates very rapidly, infiltrating neighboring tissues and spreading to lymph nodes and distant organs, such as lungs, bones, bone marrow, central nervous system, liver and retroperitoneal space. Metastases to distant organs, especially lungs, occur statistically more often than to lymph nodes [8]. The outcome of PM RMS is still unsatisfactory compared to other sites, necessitating a more aggressive therapy mostly due to delay in diagnosis [9].

The treatment of RMS involves a multimodal therapy. Treatment should be undertaken in oncology centers with multidisciplinary teams. The treatment involves surgery, chemotherapy and adjuvants and/or neoadjuvant radiotherapy . Initial surgery can be considered in patients with low risk localized embryonal non-orbital non-parameningeal RMS ; Radiotherapy is needed for satisfactory local control .Both Radiotherapy chemotherapy improves the prognosis [10].

Conclusion

The nasopharyngeal rhabdomyosarcomas are rare and their prognosis remain unfavorable. Here, we present a case of an embryonal nasophrayngeal RMS that causes to the patient an OAS and peripheral facial palsy that can be explained by her cranial extension and agressivity.

Competing Interests

The authors declare no competing interests.

Authors' Contributions

All the authors have read and agreed to the final manuscript.

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