RECURRENT EMBRYONAL RHABDOMYOSARCOMA OF THE NASOPHARYNX: A CASE REPORT

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ABSTRACT

Rhabdomyosarcoma is a malignant tumor of skeletal muscle origin. The areas of the body most commonly involved are the head, neck, orbits, genitourinary tract and extremities. Embryonal Rhabdomyosarcoma is the most common histologic variant which commonly affects children between the ages of 0 to 4 years. One subtype of Embryonal Rhabdomyosarcoma is Botryoid Embryonal Rhabdomyosarcoma which involves the mucosal linings of the nasopharynx, vagina and bladder. The cases where the nasopharynx is involved are mostly reported in children. In this case report the case of a 10-year-old boy been presented who reported to the hospital with right nasal swelling, obstruction, occasional bleeding from the nose and eye involvement. He was first diagnosed as a case of Embryonal Rhabdomyosarcoma before a final diagnosis of Embryonal Rhabdomyosarcoma of the Nasopharynx was made. He was treated with a combination of chemotherapy, radiation therapy and surgery. This case report demonstrates the recurrent nature and rare site of origin of the Embryonal Rhabdomyosarcoma of the Nasopharynx.

Key Words: Rhabdomyosarcoma, Nasopharynx, Botryoid, Embryonal.

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INTRODUCTION

Rhabdomyosarcoma, is a neoplasm of Skeletal muscle origin and has malignant potential [1]. Rhabdomyosarcoma accounts for 15 -20 % of all soft tissue sarcomas in children [2]. It affects males more than the females [3]. The most common site for occurrence in children is the orbit. There were very few cases of Rhabdomyosarcoma that had involvement of the nasal cavity [4]. Nasopharyngeal Embryonal Rhabdomyosarcoma is a rare disease. According to a study conducted in 1976, sixteen out of the two hundred and fifty-six non-epithelial lesions of the area of the nose, PNS and Nasopharynx were Rhabdomyosarcoma and out of these 16 cases only 3 revealed an involvement of the Nasopharynx [5]. The purpose of this case report was to report the case of a 10-year-old boy with Recurrent Nasopharyngeal Carcinoma and discuss its clinical, histopathological and radio-graphical findings.

CASE REPORT

A 10-year-old male patient presented to the OPD of Children hospital, Lahore, with a swelling on the right side of his nose (figure-1). His medical history revealed that he had undergone nasal surgery from Civil Hospital, Sheikhupura. He gave a history of nasal obstruction and occasional bleeding from the site.

Figure-1: Pre-operative photograph of the patient showing swelling of the nose and involvement of the eye.

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The mass was excised and sent for histopathological examination. The histopathology report revealed the presence of polyploidal tissue fragments covered with respiratory mucosa with underlying spindle cell proliferation and myxoid stroma. Cells were racket shaped to spindle cells with eosinophilic cytoplasm, vesicular nucleus, prominent nucleoli and scanty mitosis (figure 2). Immunohistochemistry staining revealed strong positive results for Desmin and positive results for Myogenin in tumor cells. A diagnosis of Embryonal Rhabdomyosarcoma was made.

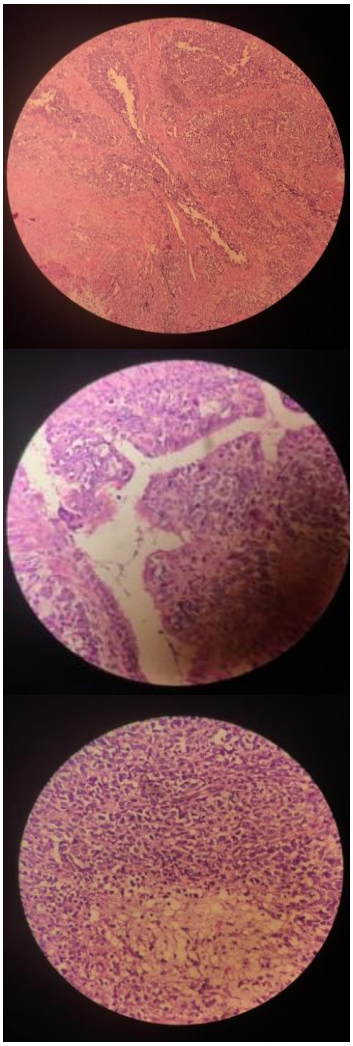

Figure-2: Histopathological findings.

The patient was then sent for CT scan of the Nose, PNS, Brain, Chest and Abdomen. The CT scan of the PNS revealed a 2×1 cm soft tissue density lesion in the left maxillary sinus along the lateral and posterior walls. Bone window images revealed no signs of bone destruction. No mass or cyst were found in the oropharynx, hypopharynx or larynx and no lymphadenopathy. The parotid and submandibular glands were normal. The patient was then started on chemotherapy. An MRI of the Nasopharynx and PNS was done three months after the Chemotherapy. Multiecho multiplanar images were performed and a 15.0 × 8.0 mm sized soft tissue density mass was seen in the left Maxillary sinus. The size of the mass had reduced from 20×10 mm to 15×8 mm compared to the pre-chemotherapy CT scan. A follow up MRI after 2 months revealed abnormal signals in the left Maxillary sinus that measured 15×8 mm and hypertrophied inferior nasal turbinates. Although the size of the left Maxillary sinus lesion was the same, there was now seen an involvement of the multiple small bilateral cervical lymph nodes. These were largest in the right upper neck measuring 16×9 mm.

The patient after two years presented with the same growth in the ENT OPD of Jinnah Hospital Lahore, in the right nasal area and reported that he had noticed it one month ago. He also reported a nasal discharge often stained with blood. He gave a history of dysphagia since the past 25 days and being on a liquid diet. Physical examination revealed a mass protruding from the right nostril stained with blood and nasal secretions and bulging of soft palate. These findings lead to a final diagnosis of Recurrent Nasopharyngeal Rhabdomyosarcoma. An MRI of the patient revealed abnormal signal intensity mass 5.1×5.2×4.5 (CC×AP×T) cm in the posterior nasal cavity and nasopharynx involving the right sided pterygoid plates and part of pterygopalatine fossa.

The preoperative workup of the patient was done. The patient was diagnosed with Hepatitis C. History of Diabetes mellitus was also revealed. The chemical pathology report revealed Serum creatinine 0.6 mg/dl, bilirubin total 0.4 mg/dl, sodium 145 mmol/l and potassium 3.9 mmol/l. The SGPT (56 U/L) and SGOT (73 U/L) were raised. Routine blood examination revealed WBC count 11.2×103 /ul, RBC count 5.29×106 /ul and Haemoglobin 13.3 g/dl. The coagulation profile of the patient was normal.

The patient was then scheduled for a surgery for the excision of the tumor. A papillomatous mass was removed in 4 to 5 isolated parts attached to right lateral nasal wall. Very little bleeding occurred. The nose was cleared completely till the right Choana.

DISCUSSION

Rhabdomyosarcoma has three main types which include Embryonal, Alveolar and Pleomorphic. The type most common in children is Embryonal. The Embryonal type is further classified into botryoid and spindle cell variants [7,8]**.** The type of Rhabdomyo-sarconma originating in the Nasopharynx, common bile duct, urinary bladder and vagina is known as Botryoid Rhabdomyosarcoma. It has a predilection for younger age group [9]. In our case the patient was

10 years old when first diagnosed with Embryonal Rhabdomyosarcoma**.**

The signs and symptoms vary greatly according to the site of involvement [10]. Rhabdomyosarcoma occurs most commonly in the head and neck region and mostly has the involvement of eye [6]. When there is involvement of the nasal cavity the patients have been seen to suffer from nasal obstruction and otitis media [4]. Our patient had an involvement of the orbit which gave his face an asymmetric appearance. Drooping of the eyelid of the involved eye was also seen. The patient also experienced nasal obstruction as there was involvement of the nasopharynx and mass was seen protruding from the nasal cavity.

The histopathological analysis is an important diagnostic modality for Rhabdomyosarcoma [11] Previous case studies have reported the presence of normal stratified squamous surface epithelium with tumor masses in the connective tissue. These tumor masses comprised of spindle and round cells with dark staining nuclei and little cytoplasm arranged in abundant myxoid stroma [12]. In our case the histopathology report revealed the presence of polyploidal tissue fragments covered with respiratory mucosa with underlying spindle cell proliferation and myxoid stroma. Cells were racket shaped to spindle cells with eosinophilic cytoplasm, vesicular nucleus, prominent nucleoli and scanty mitosis.

According to previous studies and cases immunohistochemical staining for Desmin, myo d1, myogenein, cd 56 and actin show positive results [2]. In the present case immunohistochemistry showed strong positive results for Desmin and positive for Myogenin. According to one case report series, the survival rate for patients with this tumor ranged from 20 to 35% [13]. Literature supports that although the primary treatment modality for this tumor is surgical excision, a combination of chemotherapy, radiation therapy and surgery have shown to increase the survival rates [14]. The patient in the presented case was also treated using the same combined modality treatment.

CONCLUSION

Nasopharyngeal Embryonal Rhabdomyosarcoma is a rare disease. In this case based on the patient`s clinical presentation and histopathological analysis an initial diagnosis of Embryonal Rhabdomyosarcoma was made and the patient even after being subjected to chemotherapy, although, demonstrated a decrease in size of the lesion but it was not eliminated completely. A final diagnosis of Recurrent Embryonal Rhabdomyosarcoma of the Nasopharynx was made two years later when the patient presented again with a larger mass in the same location and with more severe symptoms. It is of immense importance that in order to salvage the patient life and functionality, a timely and appropriate diagnosis of such rare cases is made and appropriate treatment modalities are employed.

AUTHORS CONTRIBUTION

Hira Butt: Manuscript writing and literature review
Nauman Rauf Khan: Manuscript writing and review
Shoaib Nasar Malhi: Treatment of patient and case monitoring
Maila Habib Piracha: Literature review
Fizza Tahir: Literature review

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