Rare Adult Sinonasal Embryonal Rhabdomyosarcoma with Optic Involvement

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Abstract

Rhabdomyosarcoma of the Paranasal sinus accounts for 10% to 15% of adult head and neck Rhabdomyosarcoma. We presented a 32-year-old man with a month history of the right eye vision loss and had no common symptoms as nasal obstruction or headache. The patient was referred to Ear Nose Throat (ENT) department, because the complaints did not improve with medical treatment. The medical history was otherwise unremarkable. The nasal endoscopic examination revealed the presence of a homogeneous, blue-purple colour and well vascularised mass and the mass filled out the right middle meatus and pushed the middle turbinate medially. There was proptosis in the right side, but there was no restriction of eye movements. The retinal posterior pole of the right eye rose, and optic disc elevated in ophthalmoscopic examination. Visual acuity was assessed as counting fingers at 20 cm in the right eye. There was no palpable lymph node in neck examination.

Navigation-assisted endoscopic complete excision of the tumour was done for surgical treatment. Immuno-histochemical examination of the mass showed strongly positive desmin (Figure 3) and positive myogenin but negative S-100, chromogranin and CD-99. It was reported as embryonal RMS.

Discussion

RMS is a fast growing, highly malignant and the most common sarcoma in childhood ages. RMS grows from mesenchymal tissue and accounts nearly for 5% to 10% of all childhood malignancies [1]. RMS is less frequently in adults than childhood [1]. There are some different clinical features between adult and child patients of RMS. Pediatric Rhabdomyosarcoma show a male preponderance, but it is not noted in adult patients. Our case was adult and RMS was located in the head and neck region but mostly adult RMS occurs in the extremities, while pediatric rhabdomyosarcoma occur predominantly in the head and neck sites [1]. RMS arises in the head and neck region in 40% of the cases, but it is less with a rate of 15% in adult RMS [2,3]. RMS of the paranasal sinus accounts for 10% to 15% of adult head and neck RMS.

Keywords: Rhabdomyosarcoma; Adult; Paranasal sinus; Vision loss

Introduction

Rhabdomyosarcoma (RMS) is a fast growing, highly malignant and the most common sarcoma in childhood ages. RMS grows from mesenchymal tissue and RMS of the Paranasal sinus accounts for 10% to 15% of adult head and neck RMS [1,2]. Nasal obstruction, pain, proptosis, limitation of eye movement and vision loss are the common symptoms but, only eye symptoms without nasal symptoms are observed less often depending on the involvement region [3]. Radiotherapy and chemotherapy is preferred with a combination of surgery for treatment [4].

Case Report

32-year-old man was presented with a history of the right eye vision loss, and the patient was followed by an ophthalmologist with diagnosis of the right eye optic papillitis. The patient was referred to Ear Nose Throat (ENT) department, because the complaints did not improve with medical treatment. The medical history was otherwise unremarkable. The nasal endoscopic examination revealed the presence of a homogeneous, blue-purple colour and well vascularised mass and the mass filled out the right middle meatus and pushed the middle turbinate medially. There was proptosis in the right side, but there was no restriction of eye movements. The retinal posterior pole of the right eye rose, and optic disc elevated in ophthalmoscopic examination. Visual acuity was assessed as counting fingers at 20 cm in the right eye. There was no palpable lymph node in neck examination.

A Computed Tomography (CT) scans of the paranasal sinus showed a “homogeneous soft” tissue mass without calcification in the right middle-upper meatus, and maxillary sinus. There was minimal damage at medial wall of right orbit (Figure 1). The mass caused a deletion between extraconal and muscle plans through right orbital apex localization, but no destruction was observed at ethmoid apex on Magnetic Resonance Images (MRI) (Figure 2).

Navigation-assisted endoscopic complete excision of the tumour was done for surgical treatment. Immuno-histochemical examination of the mass showed strongly positive desmin (Figure 3) and positive myogenin but negative S-100, chromogranin and CD-99. It was reported as embryonal RMS.

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endoscopic evaluation should be done in terms of differential and early diagnosis.

Subtypes of RMS are embryonal (70%), alveolar (20%) and other subtypes (10%). Alveolar subtype has the worst prognosis [5]. Alveolar rhabdomyosarcoma (ARMS) has a distinct histological appearance and usually presents in the soft tissues of the extremities, but Embryonal Rhabdomyosarcoma (ERMS) is often associated with mucosal or epithelial surfaces. ERMS is particularly found in the biliary system, vagina, head and neck, parametrical region and urinaiy bladder. Pleomorphic RMS is more often encountered in adults than childhood. Our case was embryonal subtype. In sinonasal tract RMS is predominantly alveolar type [2,5] and ERMS is rare and our case was an adult ethmoidal ERMS. Ahmed and Tsokos [2] reported 14 sinonasal RMS with an age range of 9 to 40 and only one patient was left maxillary ERMS and others were ARMS. Another study reported 12 adult sinonasal RMS and 2 of them were ERMS and among ERMS, one patient had an ethmoid involving as in our case [5].

CT and MRI should be done to assess the spread of the disease and the relationship between the neighbourhoud structures. CT shows the margins and relation with vital structures of masses. MRI provides superior soft tissue resolution and has high sensitivity in the pterygopalatine fossa, sinuses, and the infra temporal fossa for tumour extension. There is a high rate of distant metastasis in adults’ more than pediatric groups, so all-body Positron Emission Tomography (PET) scanning is recommended. Our case had no distant metastases in PET-CT scan.

Endoscopy and biopsy are required to confirm the diagnosis and then pathological examination gives definitive diagnosis. There are not any pathognomonic endoscopic findings in RMS. Small paranasal sinus masses could not be identified endoscopically. Desmin and myogenin is painted as positive but cytokeratin, epithelial membrane antigen, CD45, CD99, S-100 are painted as negative in immunohistochemical examination [2,6]. The pattern of immunostaining for myogenin has been used to aid in the distinction of ARMS from ERMS [5]. Embryonal tumours characteristically show myogenin in < 50% of cells as in our case. We did not perform cytophenetic and RT-PCR analysis because the tumour cells are well differentiated and we gave careful attention to use histological ancillary techniques and immunostains to clarify this differential diagnosis. This undifferentiated histology can mimic other round-cell tumours, such as lymphoma, Ewing sarcoma, and olfactory neuroblastoma [7].

Ectopic pituitary adenoma, metastatic undifferentiated carcinoma, inflammatory nasal polypsis, inverted papilloma, schwannoma, pleomorphic adenoma, squamous cell carcinoma, lymphoma, osteogenic sarcoma, fibrosarcoma, chondrosarcoma and olfactory neuroblastoma might be considered in the differential diagnosis in sinonasal cavity masses [7]. Although RMS is distinctly rare in the sinonasal tract in adults, it should always be a consideration when a poorly differentiated malignant neoplasm is presented in sinonasal region regardless of the age of the patient.

The prognosis depends on the primary site, the histological subtype and tumour size [8]. Good prognosis for survival are; tumour to be smaller than 5 cm, the patients under 20 years of age, lack of regional or distant metastasis and negative surgical margins [6,9]. Tumours of the orbit have a better prognosis. Parameningeal RMS has more recurrence rates and has much more early metastasis because of the possibility of intracerebral spread [8]. Distant metastasis occurs in adults at the time of diagnosis in 60% of cases [1]. The 5-year survival rates are 56%

Figure 2: Magnetic resonance image (Magnetom Vision; Siemens Medical Systems, PA, USA). The mass caused a deletion between extraconal and muscle plans through right orbital apex localization (Image was performed on a Gadolinium enhanced 1, 5 -T system).

Figure 3: Immunohistochemical examination showed strongly positive desmin. (X40 magnification and immunohistochemical staining).
to 65% [8]. There is not a fully-formed adult treatment protocol, but complete excision of the mass in pediatric treatment protocol is the prognostic factor that increases local control and survival [8].

Treatment of the RMS varies depending on the pathologic and clinical stage. Radiotherapy and chemotherapy are preferred with a combination of surgery [4]. Callender et al. reported 75% locoregional recurrence rate among patients with RMS after surgery [10]. Surgery is not used as a primary treatment modality [10]. The Intergroup Rhabdomyosarcoma Study Group (IRSG) recommends surgery for initial therapy, if there will be no functional loss followed by chemotherapy [11]. Patients have a better prognosis if the mass is resected completely. Residual tumour masses could be removed after chemotherapy [11]. Adult RMS is radiosensitive as other adult sarcomas, so radiotherapy is another choice of treatment modality. Occult lymph node metastases are uncommon; therefore, preventive neck dissection is not indicated. Endoscopic complete mass excision was done successfully instead of open surgery in our case, and chemotherapy was added, but radiotherapy was not preferred. No recurrence was observed during follow up for six months.

Conclusion

RMS is the most common sinonasal tumour in childhood age, but it is extremely rare in adults. Patients with sinonasal tumours often have nasal symptoms but also insidious eye symptoms could be seen rarely. If an adult patient has unilateral nasal complaint with eye symptoms, endoscopic evaluation should be done in terms of differential and early diagnosis by an otorhinolaryngologist.

References