CASE REPORT INTERNATIONAL JOURNAL OF PURE MEDICAL RESEARCH

Clinical Science

KEYWORDS: soft tissue sarcoma, nasal mass, rhabdomyosarcoma

RHABDOMYOSARCOMA OF THE PARANASAL SINUS, A RARE PRESENTATION

Volume - 5, Issue - 6, June - 2020 ISSN (O): 2618-0774 | ISSN (P): 2618-0766

Dr Bhushan Kathuria* Department of ENT/Head & Neck Oncosurgery Kainos super speciality hospital, Rohtak Haryana 124001*Corresponding Author kathuriabhushan56@gmail.com

Dr Himani Dhingra MD pediatric, DNB, FNB Pediatric Hematoncology, Senior medical officer, KEM hospital , Mumbai, Maharasthra

Dr SPS Yadav Head of department of otorhinolaryngology, PGIMS Rohtak, Haryana

Dr Deepak Nain MS , resident surgical oncology, BLK Hospital Delhi.

INTERNATIONAL JOURNAL OF PURE MEDICAL RESEARCH

Abstract

Rhabdomyosarcoma is a fast growing, highly malignant and the most common sarcoma in childhood age arises from mesenchymal tissue. Head and neck forms are not unusual, but paranasal sinus locations are very rare, that accounts for 10% to 15% of adult head and neck rhabdomyosarcoma. Here we are going to discuss about a case of geriatric maxillary rhabdomyosarcoma which was initially managed as chronic sinusitis outside, later on diagnosis was made and it was managed accordingly but prognostic outcome was very poor.1-3

Case report

A 65 year male was presented with recent episode of epistaxis from the right side nasal cavity. There was a history of using intranasal steroids, oral decongestant and oral antibiotics for 15 days as prescribed by their nearby medical practitioner in view of suspecting chronic sinusitis. There was history of on and off nasal blockage and discharge since 6 months but patient was not taking any treatment. The patient was chronic smoker, alcoholic and was wood worker as professional. General condition of the patient was not fair and was chronic hypertensive and diabetic. On evaluation, anterior rhinoscopy examination showed reddish granulomatous mass was seen near the right middle turbinate area. (fig.1) The septum was in midline. The mass was insensitive to touch and it was bleeding on manipulation. Maxillary sinus tendernes was present. Epistaxis episode was managed conservatively.

Diagnostic nasal endoscopic examination showed a red color mass protruding from right maxillary ostium, pushing the middle turbinate medially and the mass was extending posteriorly about 1cm before choana and anteriorly about 1.5 cm away from the columella.(fig.1) Left side nasal cavity was normal. Clinically we were suspecting a nasal mass with differential of inverted papilloma, sqamous cell or adenocarcinoma.

Paranasal sinuses computerized tomographic scan was showed a hyperechoic mass of size about 40 × 25 mm, filling the right maxillary sinus, with involvement of anterior group of ethmoid sinus and there was bone erosion of floor of orbit and papyraceous lamina without any intra- orbital extension. Eye examination was done that showed normal visual acuity and vision was normal. Systemic examination showed no abnormalities. The clinical history, examination and the radiological finding were in favour of carcinoma. Endoscopic punch biopsy was taken under local anaesthesia and sent for histopathological examination.

Histopathological report revealed that a tissue composed of pleomorphic polygonal atypical cells. Cells showed abundant eosinophyllic cytoplasm and vesicular to hyperchromatic nucleus with prominent eosinophilic nucleoli. Immunohistochemistry was performed that was positive for anti-desmin, anti- NCAM (neural cell adhesion molecule), anti-myogenin antibodies, and negative for anti-caldesmon antibodies.

In view of histopathological features, the diagnosis of rhabdomyosarcoma of paranasal sinus was made. The case was discussed in multidisciplinary team meeting board and it was decided to treat with primary radical surgery. But patient was very reluctant to radical surgical option and was comfortable with alternative option. So pre-treatment systemic including cardiac evaluation was done and adriamycin based single agent chemotherapy had been by started Medical Oncologist in view of performance status and multiple comorbidities. But the patient was died 12 weeks after initiation of chemotherapy that was before response assesment of the treatment. The death event was difficult to explained but may be because of chemotherapy toxicity.

Discussion

Rhabdomyosarcoma is a rare type of malignancy but the most frequent form of soft-tissue tumor, originating in the conjunctiva and its derivative tissue. It is mainly seen in childrens whereas very rare in adults. Rhabdomyosarcoma mainly develops in the thorax or extremities in adults, whereas in the childrens the head and neck or uro-genital tract are the sites of involvement.1 As per literature search, less than 10 percent of adult Head and neck locations are in the paranasal sinuses.2 Dito and Batsakis etal in their series found that 1/3 rd of total number of cases in the orbit and only 2 cases (<2%) were in nose and paranasal sinuses.3 Similarly Horn and Enterline in a study of 30 cases found only one case involving the maxillary sinus.4 In our experience of 20 years it is the only case of RMS in about 90 cases of paranasal sinuses malignancies.

Sinus involvement usually presents as early epistaxis and rarely associated with exophthalmia and ophthalmoplegia. Imaging does not help to reach defenite diagnosis, but does help in assessment of the locoregional extension. Only histopathological examination with immunohistochemistry provides specific diagnosis.

Histologically, three types are described in various literature and these are embryonic, alveolar and pleomorphic type. The embryonic type is the one most frequently encountered in ear,nose and throat region and associated with the best prognosis. The alveolar type may be located in the sinonasal cavities, and is of poorer prognosis. The pleomorphic form is rarer, mainly affects adults and is of worst prognosis.5 The case we discussed here, was histopathologically pleomorphic form so probable prognosis and

treatment modalities were explained to the patient and caretaker.

Due to the rarity of the disease in adults, there is a lack of standardized treatment guidelines and protocol unlikely as in pediatric rhabdomyosarcoma management. One retrospective study of adult rhabdomyosarcoma patients demonstrated that those patients treated according to current pediatric guidelines had similar response rates and outcome to pediatric rhabdomyosarcoma patients, suggesting that adults may be treated with similar treatment protocols as children with rhabdomyosarcoma.[6]

Currently, multimodality treatment that includes neo-adjuvent or concurrent chemotherapy, surgery with or without radiotherapy, and single or multiple drug combination chemotherapy has become the standard of care for rhabdomyosarcoma. Whatever the treatment, prognosis in adults is poor.[7,8]

Various studies in the literature reported very less mean survival in head and neck region rhabdomyosarcoma. Mason and Soul reported a generally poor prognosis in 88 cases of rhabdomyosarcoma of which 88% died within 5 year and 55% within the Ist year.9 Whereas MacComb et al. reported the mean survival of these cases <6 months irrespective of treatment administered.10 Similarly, in our case with advanced presentation, the prognostic outcome was not favorable.

Prognosis is very poor in adults than childrens, because of advanced disease presentation that may lead it to unresectable or significant risk for functional or cosmetic morbidity, also absence of standardized protocols for adult patients, and low inclusion in trials for adolescents and young adults contribute to unfavorable prognostic outcome.11

Conclusion

Rhabdomyosarcoma of paranasal sinus is very rare, but when we came across a patient with paranasal sinuses mass, we should keep in mind the possibilities of rare histopathological diagnosis including rhabdomyosarcoma; otherwise situation will lead to diagnostic dilemma and a correct diadnosis and management will be delayed

Compliance with Ethical Standards

Conflict of interest: None

Ethical issues: Informed written consent has been obtained from the patient.

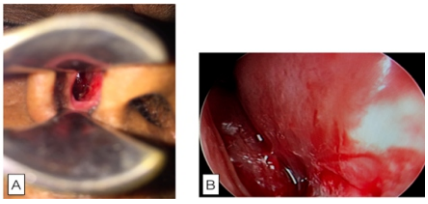

Figure 1 : Anterior rhinoscopy examination showing reddish mass near right middle turbinate area [B] Diagnostic nasal endoscopic findings .

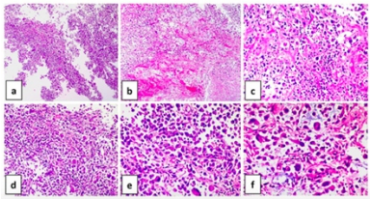

Figure 2: Histopathological examination of sinus Rhabdomyosarcoma. Chorion infiltration by loosely arranged round-cell proliferation. Numerous intra-tumoral cytonuclear abnormalities and mitoses. Dense eosinophilic aspect of the cytoplasm of certain tumor cells, suggestive of rhabdoid differentiation.

REFERENCES

1. Chen SC, Bee YS, Lin MC, et al. Extensive alveolar-type paranasal sinus and orbit rhabdomyosarcoma with intracranial invasion treated successfully. J Chin Med Assoc. 2011;74: 140—3.
2. Callender TA, Weber RS, Janjan N, et al. Rhabdomyosarcoma of the nose and paranasal sinuses in adults and children. Otolaryngol Head Neck Surg. 1995;112:252—7.
3. Dito WR and Batsakis JG : Rhabdomyosarcoma of head and neck : Appraisal of biologic behaviour in 170 cases. Ar- chives of Surgery, 1962;84 : 582.
4. Horn, R.C Jr & Enterline, H.F. Rhabdomyosarcoma A clinicopathological study and classification of 39 cases. Cancer.1958;11:181.
5. Legendre BW, Nelson BL. Alveolar rhabdomyosarcoma of the paranasal sinuses. Head Neck Pathol 2008;2: 302—4.
6. Van Gaal JC, De Bont ES, Kaal SE, Versleijen-Jonkers Y, van der Graaf WT. Building the bridge between rhabdomyosarcoma in children, adolescents and young adults: the road ahead. Crit Rev Oncol Hematol.2012;82(3):259–79.
7. Donaldson SS, Anderson JR. Rhabdomyosarcoma: many similarities, a few philosophical differences. J Clin Oncol.2005;23:2586—7.
8. Burke M, Anderson JR, Kao SC, et al. Assessment of response to induction therapy and its influence on 5-year failure-free survival in group III rhabdomyosarcoma: the Intergroup Rhab- domyosarcoma Study-IV experience —a report from the Soft Tissue Sarcoma Committee of the Children's Oncology Group. J Clin Oncol. 2007;25:4909—13.
9. Mason J.K. & Soule, E.H. Embryonal rhabdomyosar- coma of head and neck : American Jounal of Surgery.1965;100,4:7.
10. MacComb, W.S. Bardwill, J.M. Butter, J.J. & Smith, J.L.Miscellaneous Tumours : Cancer of the head and neck, Journal of the American Medical Association.1967;137 :1366 :6
11. Moon HS, Kwon SW, Lee JH. A case of alveolar rhabdomyosar- coma of the ethmoid sinus invading the orbit in an adult. Korean J Ophthalmol. 2006;20:70—5.