A Case of Rhabdomyosarcoma of the Sino Nasal Tract in an Adult Male: A Rare Case Report with Review of Literature

Ajay Manickam*, Mukesh Kumar Singh, Shaswati Sengupta, IN Kundu, Jayanta Saha, Basu SK and Rajarshi Sannigrahi

Department of ENT and Head Neck Surgery, RG Kar Medical College and Hospital, Kolkata, India

Abstract

Soft tissue sarcomas of head and neck are very rare, in adult population, that too a peripheral nerve sheath tumour getting transformed slowly on a due course of time to rhabdomyosarcomas is very rare. When we are encountering a patient with peripheral nerve sheath tumour, the possibility about malignant transformation to a soft tissue sarcoma must be explained to the patient and should be on regular follow-up.

Keywords: Rhabdomyosarcomas; Nasal cavity

Introduction

Rhabdomyosarcomas are very common soft tissue sarcomas in paediatric age group. They constitute about 4-5% of all childhood malignancies [1]. Adult rhabdomyosarcomas are relatively very rare. Adult cases are usually found to have a very bad prognosis. This is condition that requires a multimodality approach to improve the cure rate and overall outcome of the patient. Sarcoma being a feature of an inherited disease is a very rare phenomenon.

This is a rare case report of an adult patient who earlier had a peripheral nerve sheath tumour excised, and after a period of 9 years slowly it progressed to a rhabdomyosarcoma.

Case Report

A 47 year old male patient presented to the department of ENT RG Kar medical college hospital with complaints of mass in the nasal cavity for the past nine years, nasal obstruction and epiphora for the past seven years. The patient was apparently well 9 years back and then he developed complaints of mass in the left side nasal cavity. The mass was causing nasal obstruction and epiphora two years later for which the patient consulted some other tertiary care hospital. Endoscopy assisted biopsy was planned. The biopsy reports turned out to be a case of schwannoma, then endoscopy assisted sino-nasal mass excision was done.

After this surgery patient again had recurrence of disease, as the patient was explained it was a benign condition, he was not willing for follow-up, patient habituated to live with the complaints of nasal obstruction. One and a half years earlier, patient started noticing that, the mass in the nasal cavity started to grow and finally protruded out of nasal cavity. Patient also had recurrent episodes of scanty epistaxis. CT scan of para nasal sinuses was done, which showed a mass in the left side maxillary antrum involving the whole nasal cavity. Endoscopy assisted biopsy was planned and reports turned out to be a case of rhabdomyosarcoma. As the tumour was limited to the sino-nasal tract total maxillectomy was planned, surgery was done and HPE reports confirmed the mass to be rhabdomyosarcoma and the patient was referred to radiotherapy department for further management. Chemotherapy was started for the patient. In spite of multimodality approach the patients general condition was getting poorer day by day and finally the patient expired after a followup of about 1 year.

Discussion

Soft tissue sarcoma of the head and neck in an adult patient is very rare. The incidence of sarcoma of Sino nasal tract is only 30% [1]. This is said to be still more uncommon because of the transformation of schwannoma to a soft tissue sarcoma. This transformation is rarely reported in literature.

The aetiology of soft tissue sarcomas can be (1) Li-Fraumeni syndrome (2) p53 mutation (3) patients with neurofibromatosis type 1 has a 5-10 % increase in the risk of soft tissue sarcoma especially malignant peripheral nerve sheath tumours [2].

In our case it can be well noticed that aetiology was, the peripheral nerve sheath tumour slowly getting transformed to rhabdomyosarcoma. As it was a case of rhabdomyosarcoma, the most common organ of metastasis is lung; CT scan of PNS was done along with HRCT thorax, to rule out any metastasis. FNAC is of very limited value in cases of soft tissue sarcoma [3].

*Corresponding author: Ajay Manickam, Department of ENT and Head Neck Surgery, RG Kar Medical College and Hospital, Kolkata, India, Tel: 91-33-2416950; E-mail: ajaymanickam87@gmail.com

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rhabdomyosarcomas usually have a very bad prognosis, and the patient died after a period of one year.

Thus points of interest to be kept in mind in this case is, (1) when the histopathological reports in an adult patient with Sino nasal mass is found to be peripheral nerve sheath tumour chances of slow transformation of this tumour into a soft tissue sarcoma though rare is possible and hence patient has to be kept on regular follow-up. (2) Earlier diagnosis is very essential for the treatment of rhabdomyosarcoma. (3) Multi-modality treatment should be motivated as early as possible and even with proper care prognosis of the condition is poor in cases of adults.

Conclusion

Soft tissue sarcomas of head and neck are very rare, in adult population, that too a peripheral nerve sheath tumour getting transformed slowly on a due course of time to rhabdomyosarcomas is very rare. When we are encountering a patient with peripheral nerve sheath tumour, the possibility about malignant transformation to a soft tissue sarcoma must be explained to the patient and should be kept on regular follow-up. Multi-modality treatment has to be motivated as early as possible.

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Figure 2: HPE showing Rhabdomyosarcoma with aplasia.

The principle surgical modality in soft tissue sarcomas were first described by Simon and Enneking [4] and it remains as the basis of sarcoma management. A wide margin excision is advised as anything less is associated with a significant rate of relapse [5].

Adult type soft tissue sarcomas are usually radio resistant tumours. In current practice adjuvant external beam radiation often follows surgery and is usual in intermediate and high grade tumour. Recently conducted studies prove that pre-operative radiotherapy followed by surgery is giving promising results [6]. But in our case the conclusive evidence was not obtained, as pathological reports were confusing, hence surgery followed by radiotherapy was planned. Following total maxillectomy the patient was immediately referred to radiotherapy department.

Haematological and cardiological workup is very essential for the initial assessment and chemotherapy follow-up. After doing surgery the patient was immediately referred to radiotherapy department for further workup. And even with a multimodality approach,