CASE REPORT

Alveolar-type rhabdomyosarcoma of nasal cavity and paranasal sinus in adult with pulmonary metastasis: A case report

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Received 24 June 2016; accepted 17 July 2016
Available online 9 August 2016

KEYWORDS
Alveolar-type rhabdomyosarcoma; Nasal cavity; Paranasal sinus

Abstract
Alveolar-type rhabdomyosarcoma of head and neck region in adult is extremely rare. We report a case of a 33-year-old lady who presented with persistent right unilateral nasal obstruction and epistaxis for one month's duration. Examination showed a fleshy, irregular-surfaced mass occupying the whole right nasal cavity and histopathological examination revealed alveolar-type rhabdomyosarcoma. Metastatic work-up showed pulmonary metastasis. After 3 months following completion of four cycles of chemotherapy, a repeated scan showed significant shrinkage of the tumor.

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1. Introduction

Rhabdomyosarcoma is a malignant tumor that originates from striated skeletal muscle, rarely occurring in the head and neck region.1 Alveolar-type rhabdomyosarcoma, which is the uncommon form, is more often encountered in teens and young adults. Due to its aggressive growth and its deep-seated locations, alveolar rhabdomyosarcomas are often harder to treat effectively in adults, thus associated with a poor prognosis.

2. Case report

A 33-year-old lady presented with right unilateral nasal obstruction and epistaxis for one month. She denied other significant history. Nasendoscopy revealed a fleshy, irregular mass occupied the whole right nasal cavity (Fig. 1). There were no palpable neck nodes. Contrast-enhanced imaging study revealed a heterogeneous enhancing mass occupying the whole right nasal cavity with lateral extension into the right maxillary sinus. It extended posterosuperiorly to the right sphenoid sinus and posteroinferiorly into the nasopharynx (Fig. 2). There was no intracranial extension. Histopathological evaluation showed a round to polygonal shaped cells with mildly pleomorphic darkly stained nuclei. The tumor cells were stained positive for Myogenin, Desmin and CD56. Thus, it was interpreted as alveolar-type rhabdomyosarcoma. A subsequent

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Peer review under responsibility of Egyptian Society of Ear, Nose, Throat and Allied Sciences.

http://dx.doi.org/10.1016/j.ejenta.2016.07.004
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metastatic evaluation revealed pulmonary metastasis. A diagnosis of alveolar-type rhabdomyosarcoma of nasal cavity and paranasal sinus with clinical stage of T2N0M1 was made. The patient was referred to an oncology center for further management. Following the completion of four cycles of multiagent chemotherapy, a repeated scan was done during her regular monthly follow up that showed significant shrinkage of the tumor.

3. Discussion

The incidence of alveolar-type rhabdomyosarcoma is predominantly in childhood and it commonly occurs on the trunk or extremities. An adult onset rhabdomyosarcoma of nasal cavity and paranasal sinus is exceedingly rare occurrence with only several cases reported in literature. However, only one case reported with distant metastasis at time of diagnosis. In this case, during her first presentation to our center, the patient was diagnosed with alveolar-type rhabdomyosarcoma which had already metastasized to the lungs.

There are several types of rhabdomyosarcoma; the commonest type, the embryonal, accounts for about 75% and is said to have a better prognosis. The alveolar-type constitutes about 20% has a poorer prognosis. A recent study showed that the commonest rhabdomyosarcoma arising from the paranasal sinus is the alveolar-type.

There is a wide spectrum of presentation based on the involvement of the sinus or surrounding structures. In our center, nasopharyngeal carcinoma is more prevalent, which was what it was initially mistaken for. Our patient presented with only typical localized nasal symptoms such as nasal obstruction and epistaxis with no other complaints. Apart from these symptoms, patients may present with discharge, pain, and swelling. Several literature reported ophthalmic presentations such as proptosis, epiphora, diplopia, retro-orbital pain and even orbital apex syndrome.

![Image 1](endoscopic-view-right-nasal-cavity-irregular-mass.png)

**Figure 1** Endoscopic view of the right nasal cavity revealed an irregular mass occupying the whole right nasal cavity which easily bleeds on touch. S = septum, IT = right inferior turbinate, F = floor of right nasal cavity.

![Image 2](contrast-enhanced-computed-tomography.png)

**Figure 2** Contrast-enhanced (axial) computed tomography revealed a huge heterogeneously enhancing mass occupying the whole right nasal cavity with lateral extension into the right maxillary sinus posteroinferiorly to sphenoid sinus and posterosuperiorly into the nasopharynx (A, B). A coronal view (C, D) shows no intraorbital or intracranial extension.
CT and MRI are used as a guidance to confirm the location, size and infiltration of the tumor. A contrast-enhanced CT imaging is done to evaluate the primary tumor and further define the surrounding vital structures as well as to look for any bone erosion. It has been reported that tumors more than 5 cm in diameter have poorer prognosis. MRI was not done in our center due to limited resources. Other metastatic evaluation includes bone scan and abdominal ultrasound is done to determine the staging.

Diagnosing rhabdomyosarcoma remains a challenge. Histology and immunochemistry is the sole method that leads to a specific diagnosis which incorporates desmin and myogenin staining. The diagnosis of alveolar-type rhabdomyosarcoma is confirmed by microscopic visualization of small round rhabdomyoblasts arranged in nests or cards separated by connective tissue trebuculae with focal areas of alveolar architecture.

In recent years the outcome of rhabdomyosarcoma has improved due to the availability of multimodal treatment for this disease. The prognosis of this disease is dependent on the stage at which it is presented. Surgical removal of the tumor and chemotherapy or combination of both remains the mainstay treatment. A complete resection is achieved by resecting the tumor along with 0.5 cm rim of normal tissue around it. Radiotherapy is reserved for patients who develop recurrence following completion of initial treatment. The duration of the chemotherapy is determined by the stage of the disease at the time of presentation. Combinations of vincristine, actinomycin-D and cyclophosphamide (VAC) are the accepted chemotherapeutic agents. In recent years, the use of brachytherapy in head and neck tumors has become increasingly popular owing to its superiority in tissue-sparing approach.

4. Conclusion

Although alveolar-type rhabdomyosarcoma of nasal cavity and paranasal sinus in adult is extremely uncommon, it should be included in differential diagnosis. Metastasis needs to be ruled out at time of diagnosis. Early diagnosis and prompt treatment will improve outcome and increase survival rate.

References