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Case Study
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Ewing's Sarcoma Report of a Case Arising from the Nasal Septum

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Abstract

Ewing's sarcoma may arise in soft tissue or bone. It most commonly arises in the legs, pelvis, and chest wall. The cause is unknown. While Ewing's sarcoma generally occurs in a younger age group, it has been reported in adults. There have been 15 cases of Ewing's sarcoma reported involving the nasal cavity or paranasal sinuses. The treatment consists of treatment consists of multi drug chemotherapy usually following surgery and/or radiation therapy.

Keywords: Ewing's sarcoma; Trauma; Nasal septum; Sinus infections or allergies

Introduction

Ewing's sarcoma may arise in soft tissue or bone. Symptoms are quite varied but may include a mass or pain, sometimes with fever. If it arises in bone it may present as a fracture. It most commonly arises in the legs, pelvis, and chest wall. The cause is unknown. It occurs most commonly in teenagers and young adults and represents 2% of childhood cancers. It is more common in Caucasians than African Americans. The diagnosis is made by biopsy based on the histomorphologic appearance as well as immunohistochemistry. It is one of the small blue round cell tumors but typically has a clear cytoplasm on H&E staining.

Case Study

A 71 year old Hispanic woman came to the office complaining of a 3 to 4 month history of slowly increasing right nasal congestion. There were no associated symptoms; specifically no bleeding, no pain, and no rhinorrhea. There was no prior history of trauma. She did not have a history of sinus infections or allergies. On nasal sinus endoscopy there was noted to be a mass in the nasal cavity on the right between the middle turbinate and septum. It appeared to be mucosally covered and was not particularly friable. A CT scan confirmed the presence of this mass which seemed to be arising from the nasal septum. There was no other evidence of nasal or sinus disease. Specifically there was no evidence of expansion or bone erosion. She was taken to the operating room, where, under general anesthesia, during endoscopic surgery, the mass was found to be attached to the nasal septum. It was easily removed with clear margins. There was no particular extensive bleeding.

The final pathology report was Ewing's sarcoma. A PET CT scan was obtained which showed no evidence of residual disease at the primary site or anywhere else in the body. She was subsequently evaluated by Radiation Therapy and Medical Oncology. Since there was no evidence of residual disease or metastasis, it was decided that she would merely be followed with careful surveillance.

She is now 10 months post-op and has been examined monthly since surgery and there has been no evidence of residual disease or recurrence.

Discussion

While Ewing's sarcoma generally occurs in a younger age group, it has been reported in adults. There have been 15 cases of Ewing's sarcoma reported involving the nasal cavity or paranasal sinuses [1-4]. It is generally felt that the sarcoma arises from a reciprocal translocation between chromosomes 11 and 22 which fuse the EWSR1 gene of chromosome 22 to the FLI1 gene of chromosome 11 [5]. Other translocations have been reported and it is now felt that EWS/FLI serves as the master regulator.

As stated earlier the tumor most commonly occurs in the pelvis and long tubular bones. Those arising in bone are often quite painful. 30% have distant metastases at presentation.

The diagnosis is made by biopsy based on the histomorphologic appearance as well as

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immunohistochemistry. It is one of the small blue round cell tumors but typically has a clear cytoplasm on H&E staining. Glycogen in the cytoplasm can be demonstrated with a positive PAS stain. It is characteristically positive for CD99. The differential diagnosis includes other small round cell tumors such as lymphoma, rhabdomyosarcoma and Desmoplastic small round cell tumors.

In the usual presentation some kind of imaging is generally obtained. Commonly magnetic resonance imaging or computed axial tomography. In this particular case a CT scan was obtained preoperatively and a PET-CT scan obtained postoperatively.

In the more usual case, treatment consists of multi drug chemotherapy usually following surgery and/or radiation therapy [6,7]. Five year survival for localized disease is about 70% [8].

In summary, we are reporting a case of Ewing's sarcoma arising from and localized to the nasal septum.

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