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Case Report

Pseudotumor of the Maxillary Sinus in a Patient with von Willebrand's Disease

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Von Willebrand's disease is a congenital bleeding disorder that is inherited as an autosomal dominant trait. The pathogenesis of bleeding is due to the absence of platelet adhesion to subendothelium that is mediated by von Willebrand's factor. The plasma of the patient is also deficient in factor VIII coagulant activity.

Hemophilic pseudotumor is an uncommon, severe complication of a bleeding diathesis that occurs in bones and soft tissues [1]. It is estimated to occur in 1–2% of those affected by bleeding diathesis and has mostly been reported in severe hemophilia [2]. When hemophilic pseudotumor involves bone, it is also called hemophilic pseudocyst, an extraarticular destructive osseous lesion causing necrosis of the bone by the hematoma under pressure. A review of the literature reveals that, to date, nine cases have been reported in the mandible and none in the maxilla, all occurring in hemophilia [2]. Hemophilic pseudocyst of the maxilla has not been documented in von Willebrand's disease to our knowledge.

We present a unique case of pseudotumor of the maxillary sinus in a patient with von Willebrand's disease.

Case Report

An 11-year-old male with a diagnosis of type 3 von Willebrand's disease was admitted to a dental clinic with complaints of a toothache in the right upper jaw, severe headache, and occasional epistaxis. Dental examination was normal, and antibiotic treatment was begun with a provisional diagnosis of acute sinusitis. His pain continued to increase despite therapy, and he was referred to our medical center for further evaluation.

On physical examination, we found the right maxillary region tender; there was slight edema of the right eyelid and mild right-sided proptosis with normal vision and eye movement. No history of trauma was obtained. Rhinoscopic examination revealed edematous right inferior concha and a blood clot in the right nasal cavity. Radiography in Waters' projection (Fig. 1A) revealed opacification of the right maxillary sinus with expansion and partial nonvisualization of its walls. A CT scan (Fig. 1B) revealed a heterogeneous soft-tissue mass that contained areas of high density and obliterated the right maxillary sinus. Upward expansion of the orbital floor was present, and the lesion extended to the posterior border of the optic bulb, filling most of the retrobulbar space and causing proptosis. The bony margins of the maxillary and ethmoid sinuses were expanded and partially destroyed, allowing the lesion to extend through these bony defects to the nasal cavity anteriorly to the subcutaneous fat tissue of the cheek and inferiorly to the root of the canine tooth. There was no enhancement on the CT scan after IV injection of contrast material (Fig. 1C).

We performed a Caldwell-Luc operation and observed that the maxillary sinus was full of organized hematoma with hyperplastic mucosa. The histopathologic examination showed an organizing hematoma with no evidence of malignancy. MR imaging after the removal of the nasal pack showed a heterogeneous soft-tissue mass much smaller than the one seen before surgery (Fig. 1D and 1E). The mass represented different phases of blood products that indicated bleeding back into the sinus. We continued managing the patient with replacement therapy.

Discussion

In bleeding disorders, the pseudotumor consists of an encapsulated blood collection caused by recurrent hemorrhage in either bone or soft tissues. Increasing pressure within the hematoma leads to erosion of adjacent tissues [1]. These lesions primarily involve the long bones of the lower extremity and pelvis. Other less common sites include the

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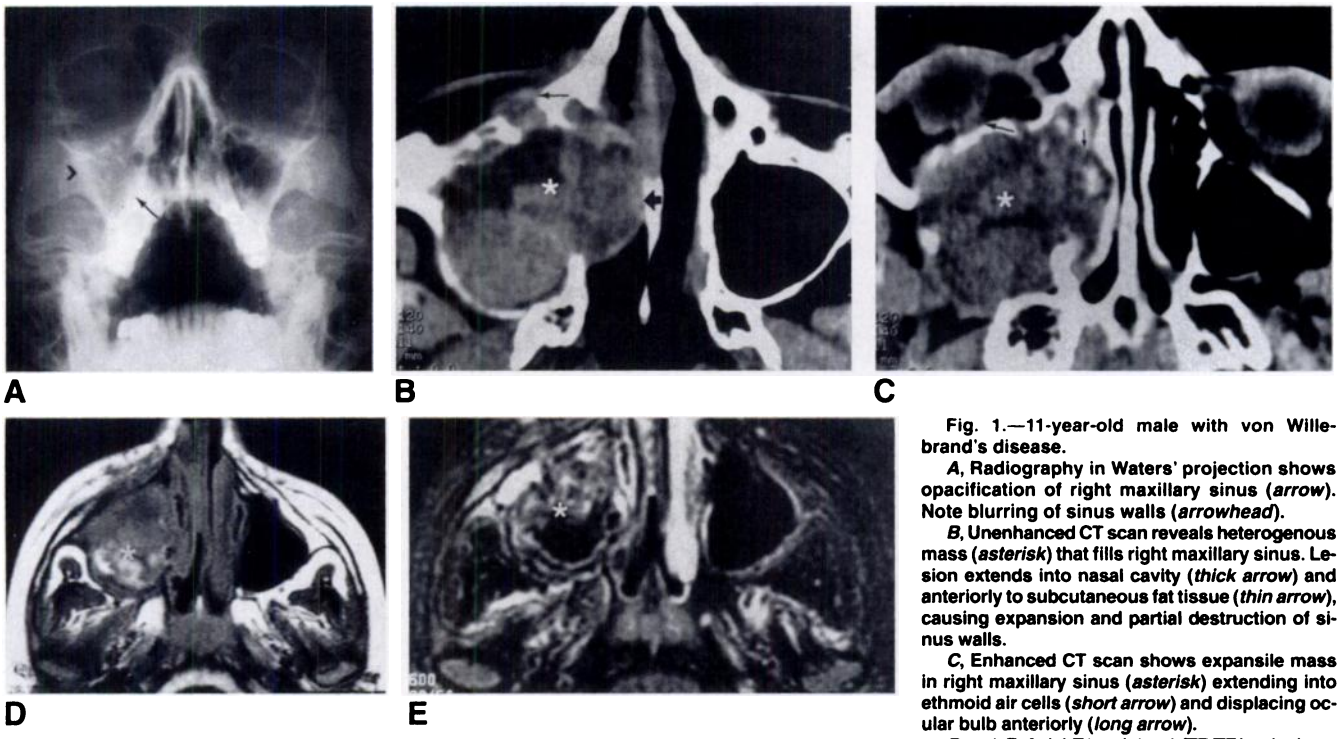


Fig. 1.—11-year-old male with von Willebrand's disease.

***A*, Radiography in Waters' projection shows opacification of right maxillary sinus (*arrow*). Note blurring of sinus walls (*arrowhead*).**

***B*, Unenhanced CT scan reveals heterogenous mass (*asterisk*) that fills right maxillary sinus. Lesion extends into nasal cavity (*thick arrow*) and anteriorly to subcutaneous fat tissue (*thin arrow*), causing expansion and partial destruction of sinus walls.**

***C*, Enhanced CT scan shows expansile mass in right maxillary sinus (*asterisk*) extending into ethmoid air cells (*short arrow*) and displacing ocular bulb anteriorly (*long arrow*).**

***D* and *E*, Axial T1-weighted (TR/TE/excitations, 500/23/4) (*D*) and T2-weighted (6600/120/3) (*E*) MR images show heterogenous signal intensity (*asterisk*) that represents different phases of blood products caused by bleeding back into sinus.**

orbits, the mandible, the clavicle, the calcaneus, the skull, the small bones of the hand, and the spinal canal [3, 4].

Pathologic studies of pseudotumors find a thick fibrous capsule surrounding a coagulum. It is thought that recurrent bleeding into this capsule, which prohibits reabsorption of the blood, accounts for the expanding nature of the lesion and the subsequent destruction of surrounding tissues.

Intraosseous hemorrhage causes a lytic bone lesion that is round in shape with sclerosis that mimics a benign tumor. Irregular contours and bony erosion may also be observed, raising the suspicion of malignancy, which was the case in our patient [5]. The differential diagnosis in our patient included neoplastic mass lesions such as ameloblastoma, esthesioneuroblastoma, squamous cell carcinoma, adenocystic carcinoma, and lymphoma. With these lesions, various degrees of contrast enhancement are expected. Rather than expanding, most of these lesions cause bone destruction. Aneurysmal bone cyst, mucocele, and odontogenic cysts were also considered in our patient [6]. Because the lesion showed no contrast enhancement and contained areas of high density, a pseudotumor was considered in the initial diagnosis, but other causes could not be excluded because of the unusual location.

Several therapeutic approaches have been used in managing hemophilic pseudotumors. Surgery, factor VIII replacement therapy, and local irradiation, alone or in combination, as well as conservative measures, have been most often described [1]. In our patient, we performed surgical excision and replacement therapy. The histopathologic examination supported our diagnosis of pseudotumor.

A provisional diagnosis of pseudotumor should be considered when a patient with severe hemophilia develops a slow-growing mass [6]. CT remains the primary diagnostic technique in such cases. MR imaging is also valuable to reveal the presence of blood products and the extension into surrounding tissues.

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