Nasal Hemophilic Pseudotumor: Favorable Response to Radiotherapy

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Hemophilic pseudotumors are rare but dangerous complications of Hemophilia. We hereby report a 3-year-old boy with Hemophilia B, presenting with nasal pseudotumor, showing favorable response to radiotherapy after unsuccessful treatment with factor IX replacement therapy. The diagnosis and treatment of this rare condition is also reviewed.

Key words: *Hemophilia, Nasal, Pseudotumor, Radiotherapy.*

Hemophilia A and B are the only two heritable bleeding disorders inherited as X-linked recessive pattern affecting exclusively the male while females are carriers. Hemophilia A (Factor VIII deficiency) is more common than hemophilia B (Factor IX deficiency). The degree of severity of clinical manifestations depends on factor level. The joints are the most frequent site of bleeding followed by the soft tissues and bones [1]. Occasionally, it presents as a pseudotumor that most commonly develops in the femur, tibia and pelvic bones, while orbit, small bones of the hand, mandible, clavicle, spinal canal are less common sites [2]. However occurrence of nasal hemophilic pseudotumor is an extremely rare and there have been only few such reports in world literature but none associated with hemophilia B [3,4].

We report here a nasal pseudotumor in a boy with Hemophilia B that presented as epistaxis with progressively increasing respiratory distress to highlight the relatively unusual location and radiotherapy as an effective modality of treatment.

CASE REPORT

A 3-year-old boy with hemophilia B was admitted with a history of progressively increasing nasal mass and intermittent epistaxis for last 1 month. Careful history revealed that about 2 months back the child had a minor trauma to the nose, followed by epistaxis, which was initially controlled over 5 days with pressure bandage and factor IX replacement. Physical examination revealed a large swelling (5X6 cm) over anterior and upper part of the nose, completely altering the normal architecture of the nose. The swelling was tense, glistening, of heterogeneous consistency and was only mildly tender to touch (***Fig.* 1**). Anterior rhinoscopy did not provide much information other than documenting the presence of the mass.

Laboratory investigations were as follows: Hemoglobin: 10 gm%, prothrombin time (PT): 12.1s (control-11.8s, INR-1.03), activated partial thromboplastin time (aPTT): 100.9s (control- 28.1s), factor VIIIc assay: 76% (ref. range: 50-150%) and factor IX assay: less than 1 % (ref. range: 50-150%).

Factor replenishment was started as an initial treatment but the condition failed to improve much and epistaxis continued, even after repeated transfusions. Moreover the size progressively increased over the next few days causing respiratory discomfort. A differential diagnosis of nasopharyngeal angiofibroma was made. Non-contrast computed tomography (CT) scan of nose and paranasal air sinuses demonstrated an externally protruding large (3.5×5 cm) soft tissue mass with high density (+70HU) involving the anterior third of both the nasal fossae with erosion of the nasal septum and smooth scalloping of the adjoining bones due to chronic pressure changes, which showed mild enhancement on contrast study, suggestive of a pseudotumor or blood cyst (***Fig.* 2**). Surgical intervention was planned but his parents refused that due to operative risks.

The child was therefore referred to the department of Radiation Oncology and external beam radiation therapy was then instituted. A total dose of 900 cGy over 6 fractions in 6 days was given by 6 MV linear accelerator. The size of pseudotumor gradually decreased, and epistaxis completely stopped by two weeks after completion of radiotherapy. The patient has been under follow-up for last six months since treatment. During this time no evidence of tumor recurrence was observed.

DISCUSSION

Pseudotumors or blood cysts are rare but dangerous complications of hemophilia, occurring in 1%-2% of patients with severe forms of the disease [1]. It is essentially a chronic, slowly expanding hematoma

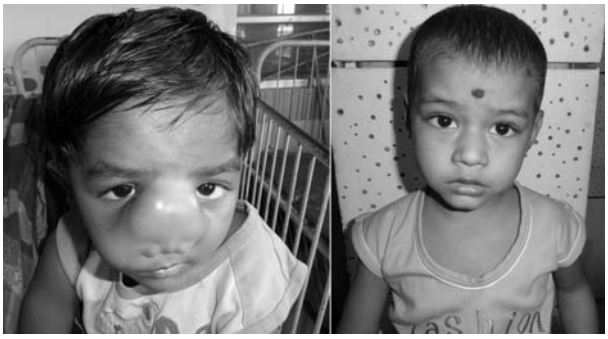

FIG.1 *A large (5×6 cms), tense, glistening swelling of heterogeneous consistency over upper part of the nose with gross narrowing of both the nostrils (left) and post-treatment (right).*

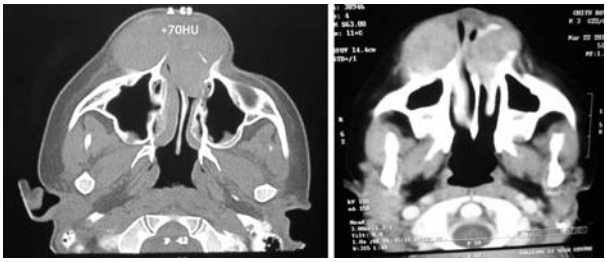

FIG.2 *Showing an externally protruding large (3.5×5 cms) soft tissue mass with high density (+ 70 HU) on plain CT with erosion of anterior nasal septum and smooth scalloping of the adjoining bones. The lesion showed mild enhancement on contrast study.*

resulting from repetitive bleeding and is surrounded by thick fibrous capsule. Many patients recall sustaining an injury prior to development of the pseudotumor [5]. As the swelling progresses, increasing pressure leads to the slow destruction of adjacent structures by progressive necrosis [6]. Invasive techniques such as, percutaneous aspiration and needle biopsies are strongly discouraged to diagnose hemophilic pseudotumor due to increased risk of complications like hemorrhage, infection and fistulization [2,6]. High quality CT scan and/or magnetic resonance imaging (MRI) is an excellent tool for preoperative visualization of the extent of the lesion, its mass effect on vital surrounding structures and possible invasion of joints. CT is particularly helpful in the evaluation of bone, whereas MRI is superior to CT for delineating soft tissue and intramedullary spaces [7].

Even though treatment of hemophilia has undergone rapid development in the past decade, at present hemophilic pseudotumor lacks standard management guidelines. Till now, the initial treatment is conservative with clotting factor replacement to keep an activity of 100%. For patient with inhibitors, recombinant factor VIIa or prothrombin complex concentrates can be used. In general, operative removal of the entire mass is a reliable treatment because the pseudotumor likely will reform if it is not completely removed [1]. Radiotherapy with or without replacement therapy has shown promising results as an alternative to a more mutilating surgery or where surgery is contraindicated, or resistant to conservative treatment. The exact mechanism of hemophilic pseudotumor to respond to radiotherapy is not known. But different opinion suggests that radiation results in: (*a*) endarteritis in an acute bleeding hematoma; (*b*) direct injury of small vessels causing fibrosis and healing; and (*c*) stimulation of fibroblasts resulting in fibrosis [8]. There has been considerable variation in the literature in radiotherapy dose. Also, the doses, as low as 600 cGy to as high as 2350 cGy, with or without factor replacement, have shown good response with complete resolution of lesions [9]. Even though, no standard radiation dose and fractionation schedule exists in the management of hemophilic pseudotumors, radiation therapy should be tried in cases where surgery is not feasible.

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