

Paranasal Sinus Involvement in Thalassemia Major: CT Demonstration

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Beta thalassemia is a blood dyscrasia caused by defective synthesis of the beta chain of hemoglobin. This causes severe anemia due to ineffective erythropoiesis and peripheral hemolysis of the defective red cells. Marked expansion of active marrow spaces and extramedullary hematopoiesis result.

Classic radiographic findings of thalassemia, in particular the hair-on-end appearance in the skull and the widening of tubular bones with a coarse trabecular pattern are well known [1]. We describe a case in which CT of the sinuses showed marked sinus and facial deformity due to marrow expansion.

Case Report

A 13-year-old girl with thalassemia major was evaluated for possible facial reconstruction for nasal obstruction that was causing respiratory difficulty. Physical examination revealed significant frontal bossing, maxillary hyperplasia, marked flattening of the nasal bridge, and enlargement of the nasal septum. There was marked protrusion of the upper teeth.

Radiographs of the skull and sinuses showed marked calvarial thickening as well as lack of pneumatization and apparent expansion of the maxillary antra (Fig. 1A). Noncontrast CT of the maxillae showed soft-tissue density in the region of the maxillary antra due to marrow overproduction in their walls, with consequent marked expansion of these structures and resultant lack of pneumatization of the air cells (Fig. 1B). The walls of the maxillary antra showed markedly expanded marrow cavities and very thin cortices, but were not destroyed. Compression of the nasal cavity, particularly on the

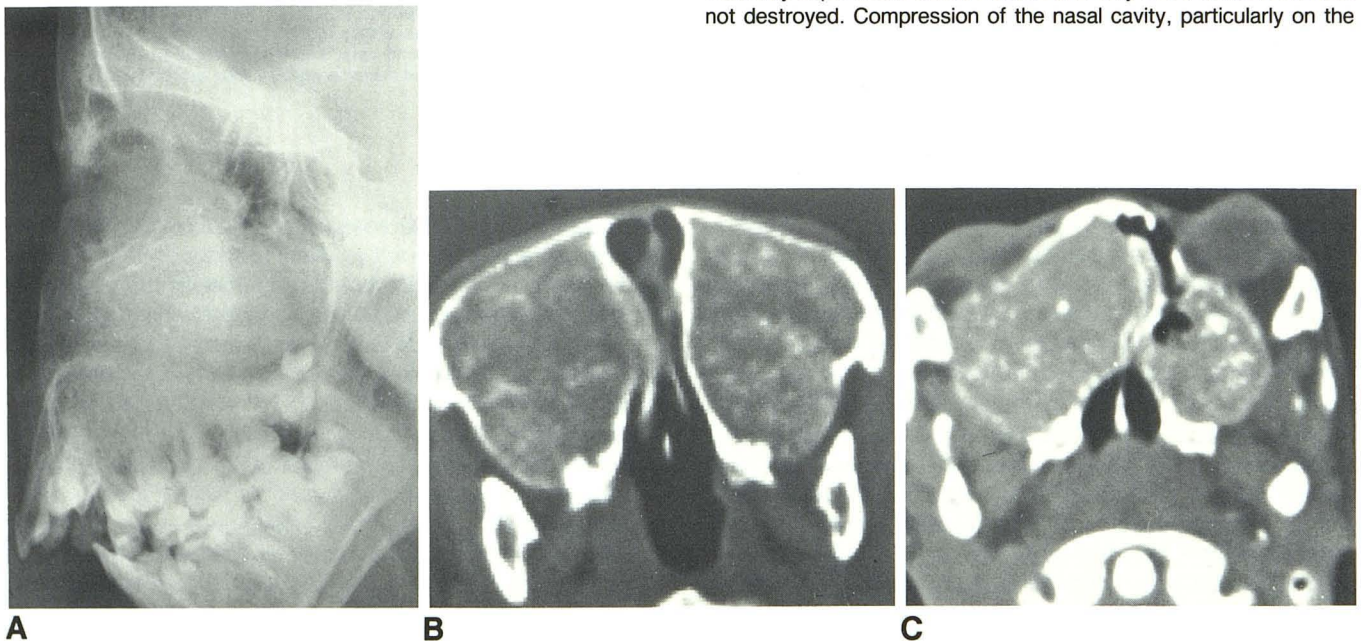


Fig. 1.—A, Lateral radiograph of face. Lack of aeration and apparent expansion of maxillary sinuses and alveolar portions of maxilla with protrusion of incisor teeth.

B, Noncontrast CT of maxilla. Thinning and expansion of antral walls by soft-tissue density with flecks of calcification due to marrow overproduction within maxilla.

C, CT at slightly higher level showing narrowing of right nasal cavity and nasal septal deviation due to expansion of right maxilla.

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right, with deviation of the nasal septum to the left was noted (Fig. 1C). Involvement of the right anterior ethmoid air cells and the lower portion of the maxilla bilaterally was noted on other images.

Discussion

Beta thalassemia is an autosomal recessive disorder that typically occurs in persons of Mediterranean origin. Patients with thalassemia major, the most severe form of the disease, are homozygous whereas the heterozygous form, thalassemia minor, is usually asymptomatic. Patients with thalassemia intermedia are homozygous, but have a milder form of the disease. They maintain hemoglobin levels above 7 g/100 ml without transfusion and have a better prognosis. Typical clinical findings include bronzing of the skin due to hemosiderosis, hepatosplenomegaly due to extramedullary hematopoiesis, and "rodent facies."

The characteristic facial changes are due to active marrow hyperplasia. Microscopic examination of the bone marrow demonstrates a variety of primitive cells, including numerous nucleated erythrocytes as well as myelocytes and megakaryocytes. The marrow expansion in the facial bones results in delay in pneumatization of the sinuses, overgrowth of the maxillae, and forward displacement of the upper incisors. Problems with malocclusion as well as cosmetic deformity occur.

CT shows soft-tissue density material in the paranasal sinuses, typically in the walls of the maxillary antra, but also involving the ethmoid sinuses. These findings are due to marrow overproduction in the maxilla. Marrow expansion in

the walls of the paranasal sinuses results in expansion of their walls with cortical thinning and impaired pneumatization. These findings have previously been described briefly [2], but have been attributed to expansion of the sinuses themselves with loss of pneumatization rather than to antral-wall expansion with delayed or impaired pneumatization. Analogous findings occur in the calvaria, where there is widening of the diploic space and thinning of the outer table.

Although it has been stated that the facial deformity in these patients can be prevented by transfusion therapy [3], they still present for correction of cosmetic and orthodontic problems. Treatment with orthodontic appliances and surgical reduction of the alveolar bone has been described [4]. Radiographs and CT provide useful information regarding deformity of the facial bones and may be extremely helpful in planning reconstructive surgery. In this case, CT clearly demonstrates the extent of involvement by delineating the amount and extent of facial deformity and encroachment on the nasal cavity and paranasal sinuses.

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