Spinal Cord Compression Secondary to Extramedullary Hematopoiesis in Two Brothers

Report of Two Cases

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Two brothers, 2 years apart, presented a picture of acute cord compression and were relieved by laminectomy and tumor excision. Both demonstrated typical paravertebral masses on plain x-ray representing rests of extramedullary blood-forming tissue, and both were diagnosed as having thalassemia intermedia.

Case Reports

Case 1. A 40-year-old Negro man, previously reported by Sorsdahl, et al., was initially admitted because of jaundice and anemia. He gave a history of having worked as a laborer until age 35 when he developed generalized weakness and palpitation. The symptoms could be relieved by frequent blood transfusions, but the patient found his exercise tolerance too low to continue work.

Examination. Physical findings included hepatosplenomegaly, poor anal sphincter tone, urinary retention, and generalized weakness. The hematocrit was 30%, and the white blood cell count 9,500 with three normoblasts per 100 white cells. The reticulocyte count was 2%, and hemoglobin electrophoresis showed 2% A2 and 23% hemoglobin F. Liver function studies were consistent with obstructive jaundice. X-rays revealed bony changes of generalized reticular appearance with scattered areas of coarse rarefaction in the skull and vertebral bodies. An intrathoracic soft tissue mass, approximately 2 × 2 cm, was found in the region of T-9 and T-10. There was no evidence of bone erosion. The patient was discharged with a diagnosis of intrathoracic extramedullary hematopoiesis secondary to thalassemia intermedia.

Second examination. Two months later the patient was readmitted for evaluation of progressive paraparesis, and urinary retention. Findings included generalized weakness in both legs, absent abdominal reflexes, and a left Babinski sign. There was decreased perception of light touch and superficial pain below the T-9 dermatome. Vibratory and position sense was also absent below this level. Thoracic spine x-rays revealed no change in the paraspinal mass. A diagnosis of spinal cord compression was made.

Operation. Emergency decompressive laminectomy was carried out from T-4 to T-8. Three discrete, posterior midline extradural masses were found to be causing cord compression and were totally removed. Subsequent microscopic examination of the surgical specimen revealed active hematopoietic tissue with numerous nucleated red cells, scattered megakaryocytes, and occasional myeloid elements.

Postoperative course. The patient had an uncomplicated postoperative course. Within 2 weeks he was walking well and was discharged. On subsequent examination, strength and sensory perception had improved markedly.

Case 2. The 47-year-old brother of the above patient was initially admitted for evaluation of anemia, which had been symptomatic since age 25.

Examination. The patient was weak and debilitated, with cardiomegaly, a systolic murmur, hepatosplenomegaly, and bilateral cryptorchidism. The hematocrit was 22% and hemoglobin 8.6 gm. The peripheral blood smear showed target cells, marked anisocytosis, a white blood cell count of 9,500,
reticulocyte count of 0.6%, and a mean corpuscular volume of 74 cu. Hemoglobin electrophoresis showed 4.6% A2 and 5.9% hemoglobin F. X-ray findings included bony changes compatible with chronic hemolytic anemia. Chest and thoracic spine x-rays showed right and left intrathoracic paraspinous masses at T-9 and T-10. The patient was discharged with a diagnosis of thalassemia intermedia.

Second examination. The patient did well until 5 months later when he developed chest pain, shortness of breath, pedal edema, and orthopnea. The physical examination showed the same findings as before. He improved after transfusion of 4 units of packed cells, and was discharged.

Third examination. Two months later, the patient was readmitted for evaluation of progressive paraparesis. The physical examination showed the additional findings of bilateral lower abdominal and leg weakness, and marked reduction in perception of all sensory modalities below the T-10 dermatome. Hyporeflexia was present. Chest x-ray showed enlargement of the paraspinous masses previously described. No overt compression of vertebral bodies or change in the posterior elements was noted. Cisternal myelogram demonstrated a complete block at T-7.

Operation. Surgical decompression exposed an extensive epidural mass of friable vascular tissue compressing the spinal cord from T-4 to T-8. The tumor was partially removed.

Microscopic examination showed it consisted of active hematopoietic tissue similar in appearance to the specimen from Case 1.

Postoperative course. The patient improved slowly and gradually returned to walking. Because of known extraspinal extension of the epidural mass, he was treated with cobalt-60 irradiation to the affected area. This patient is now doing well 2½ years after surgery.

Discussion

As previously cited, Sorsdahl, et al., reported the first of these patients in 1964. In their thorough review of the literature, they found 19 other cases of thoracic extramedullary hematopoiesis but only three cases of clinically obvious secondary spinal cord compression.

Extramedullary hematopoietic tissue may develop in quite varied sites. Although the most common sites are liver, spleen, and lymph nodes, foci of hematopoietic tissue have been reported in the brain, along peripheral nerves, and in adipose tissue. The etiology of ectopic foci of hematopoietic tissue remains obscure. The masses may arise when bone marrow potential is not adequate to meet the demands of the organism. This compensatory mechanism is seen in the hemolytic anemias, nutritional deficiency anemias, myelosclerosis, carcinomatous replacement of the marrow, leukemia, and lymphomatous diseases. There are many hypotheses concerning the origin of these hematopoietic tissue masses. Lowman, et al., suggest that they develop either from embryonal rests of osteogenic tissue or from bone marrow emboli undergoing metaplastic change to form hematopoietic tissue masses. The origin of such tissue in the paravertebral region has been suggested by Lyall as direct extension of subperiosteal marrow from adjacent vertebral marrow cavities.

The extramedullary hematopoietic tissue masses found in the spinal canals of these patients presumably developed in response to their rather severe anemia. Thalassemia intermedia is a genetically determined defect in hemoglobin synthesis resulting in defective hypochromic microcytic red cells, according to Pearson and Noyes. It occurs in three genetically determined forms: thalassemia major, the homozygous condition; thalassemia minor, the heterozygous form; and thalassemia intermedia, an interaction between thalassemia and another normal hemoglobin gene. Although the hemoglobin A2 and hemoglobin F levels in these patients follow no particular pattern (one suggesting thalassemia major, the other suggesting thalassemia minor), the diagnosis of thalassemia intermedia was made by the late onset of symptomatic anemia and by its severity, which was greater than usually seen with thalassemia minor.

Summary

We have described two cases of spinal cord compression secondary to extramedul-
Spinal Cord Compression Due to Hematopoiesis

lary hematopoiesis that were a diagnostic
challenge and particularly interesting be-
cause they occurred in brothers.

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