Eosinophilic granuloma of bone was first reported as a clinical entity in 1940 by Otani and Ehrlich and independently by Lichtenstein and Jaffe. In 1942 Green and Farber and later in 1944, Jaffe and Lichtenstein advanced the idea that Hand-Schüller-Christian’s disease, Letterer-Siwe’s disease and eosinophilic granuloma are allied, but these authors differed on the question of etiology. The present case is reported because of the unusual size of the granuloma, the presence of multiple bone lesions and the accompanying thrombocytopenic purpura, which has been reported as a feature of Letterer-Siwe’s disease in infants and children.

REPORT OF A CASE

A 57-year-old Jewish housewife was admitted to the Gallinger Municipal Hospital with a draining sinus at the vertex, of 6 months’ duration. The past history revealed a hemorrhagic tendency with excessive bleeding when cut, easy bruising and frequent nosebleeds since the age of 5. One year prior to admission a painful area developed at the vertex shortly after the patient had had several teeth extracted. This finally opened spontaneously and began to drain. At the same time a draining sinus developed on the right mandible near the chin, together with tinnitus of the left ear and tenderness over the left mastoid. The lesion at the vertex was associated with intermittent pain.

Examination revealed an ulcer at the vertex, 2.5 cm. in diameter, and an area of softening under the scalp, extending over both the posterior parietals and anterior portion of the occipital bones and measuring 15 cm. in diameter. A draining sinus was also present on the right mandible and a serosanguineous discharge came from the left auditory meatus. There were numerous petechial hemorrhages over the abdomen and areas of purpura and ecchymosis over the extremities. Neurologic findings were normal.

The initial laboratory studies revealed no abnormalities except for a platelet count of 90,000. A biopsy of the margin of the scalp lesion and underlying tissue was reported as “chronic granulation tissue.” Bone marrow studies showed an increased megakaryocyte count with a decreased platelet production. Bleeding time was prolonged. Roentgenograms revealed an extensive area of bone destruction in the skull with no new bone formation (Fig. 1).

Course. A splenectomy was done in an attempt to correct the thrombocytopenia. Six weeks later the patient’s platelet count was still only 36,000. Nevertheless, because of the tremendous size of the bony defect, surgery was decided upon. The preoperative diagnosis was “chronic osteomyelitis” of the skull.

Operation. Under endotracheal anesthesia an incision was made from ear to ear over the vertex of the skull, similar to a postmortem incision. The scalp was stripped back, exposing a large cranial defect with ragged borders and a great deal of granulomatous tissue adherent to the dura mater. This was stripped off in sheets and there was considerable bleeding. The bone, also, was very vascular and the procedure of bitting it away was accompanied by continuous blood loss. A defect which measured approximately 15 x 15 cm. was thus created. Because of the favorable reports on the use of tantalum plates for acute skull defects, we decided to insert a tantalum plate as a primary procedure in the hope that any further infec-
tion might be controlled by massive doses of antibiotics, and that the patient could weather through the procedure. Accordingly, a 15 by 15 cm. tantalum plate, neither trimmed nor otherwise altered except for perforating it and bending it to conform to the shape of the skull, was fitted in over the defect (which even now was not fully covered) and secured in place with tantalum screws. Beneath the plate a drain was placed and led out through a stab incision.

During the operation the patient received 3500 cc. of blood and lost at least an equal amount. However, upon leaving the operating room she was awake and responded slowly though her blood pressure was 70/40.

Pathological Examination. The report by the pathologist revealed for the first time that the lesion was an eosinophilic granuloma (Fig. 2).

Course. Two weeks postoperatively the original draining sinus at the vertex reopened though the initial surgical incision healed per primam. The sinus was again excised and closed 1 month later but after about 2 weeks it again broke down and drained. Shortly after the healing of the new excisions a course of deep roentgen therapy, totalling 1350 R units (450 R through three ports) was given, with a dramatic change in the X-ray picture in 1 month’s time. However, shortly after the X-ray therapy was given, again the central wound broke down. In spite of this the patient insisted on being discharged with a draining sinus at the vertex and the plate plainly visible.

Three months later she was re-admitted for removal of the plate. The initial coronal incision was reopened, the scalp was stripped back and the plate was removed from its bed. Between it and the dura mater there was a small amount of granulation tissue. The coronal incision was then closed. The site of the old draining sinus was then sutured and in accom-