Brief Communication
Communication abrégée

MASSIVE SUBCUTANEOUS HEMORRHAGE IN A CHEST-WALL NEUROFIBROMA

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Congenital neurofibromatosis type 1 (NF-1) or von Recklinghausen's disease is an autosomal dominant disorder characterized by neurofibromas, pigmented skin lesions (café-au-lait spots), iris hamartomas, meningeal tumours and, rarely, by autonomic ganglia tumours such as pheochromocytomas.[1] There have been case reports of an association between NF-1 and von Willebrand's disease,[2] characterized by attenuated platelet sensitivity to collagen.[2] This defect may predispose patients with NF-1 to prolonged bleeding after minor trauma. Life-threatening spontaneous hemorrhage in NF-1 patients has been reported, thought to be a result of friable vasculature secondary to arterial dysplasia or vascular invasion by the neurofibroma.[1] We report a case in which a patient with NF-1 presented with mild traumatic injury to a chest-wall neurofibroma that progressed to massive, life-threatening subcutaneous hemorrhage.

CASE REPORT

A 25-year-old woman with known NF-1 presented with a painful bruise in the posterior right subcostal region. The NF-1 had been diagnosed from biopy of a café-au-lait spot when she was 2 years old and she had been followed up in the neurology clinic of the Hospital for Sick Children in Toronto. She had cerebral infarcts as a child and was left with a residual right hemiparesis and epilepsy. She also had poorly controlled hypertension and an arteriovenous malformation in the left occipital region.

The patient admitted to falling from a chairlift while skiing 4 hours before presentation. Initially, there was no evidence of injury, but she noticed a gradual increase in the size of her bruise with significant swelling and tenderness. On initial physical examination, she was hemodynamically stable with a large hematoma measuring $30 \times 15 \times 10$ cm in the right flank, extending from the subcostal margin to the posterior superior iliac spine. Otherwise the physical findings were unremarkable and there was no evidence of respiratory distress or peritoneal findings suggestive of intra-abdominal bleeding. The hemoglobin level on presentation was 139 g/L with an international normalized ratio (INR) of 1.34 and a partial thromboplastin time (PTT) of 28.7 seconds. During the next 6 hours the hematoma expanded, and there was an associated fall in the hemoglobin level to 58 g/L. In addition, a bleeding diathesis occurred, with the INR rising to 3.39, the PTT to 107 seconds and a critical fall in fibrinogen to 0.6 g/L. At this point, the patient became hemodynamically unstable with a sinus tachycardia and hypotension.

The patient was resuscitated with a total of 10 units of packed red blood cells, 2 units of fresh frozen plasma, 8.5 L of normal saline, 10 units of platelets and 10 mg of vitamin K. Because of her ongoing fluid requirements and continued expansion of the hematoma, the patient was operated on to evacuate the hematoma and control the bleeding. Intraoperatively, the wound was explored, but no single bleeding source or vascular abnormality was identified. The central portion of the hematoma was necrotic and was debrided. The wound was packed open with saline-soaked gauze and she was returned to the intensive care unit in stable but critical condition.

The patient remained stable postoperatively with no evidence of further bleeding. Two days later the dressing was changed and the wound explored. During her subsequent hospital stay she required an additional 4 intraoperative wound explorations, débridement and dressing changes. A final débridement and primary wound closure were done on hospital day 26. She was discharged home in good condition 34 days after admission. During her hospitalization, she received 35 units of packed red blood cells, 22 units of fresh frozen plasma, 35 units of platelets and 20 units of cryoprecipitate.

Pathological examination of her resected wound margins revealed that the hematoma originated in a cutaneous neurofibroma which stained positive for S-100 protein and contained mast cells (Fig. 1). There was no evidence of arterial dysplasia

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Accepted for publication July 4, 1999.

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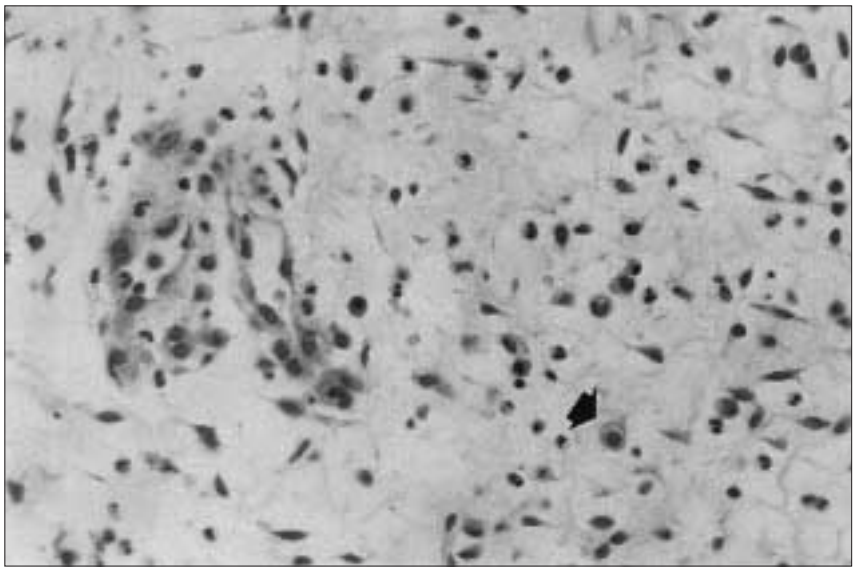

FIG. 1. Photomicrograph of the resected tissue demonstrating the mast cells (arrow). Mast-cell infiltration is a histologic characteristic of cutaneous neurofibromas (hematoxylin–eosin, original magnification × 400).

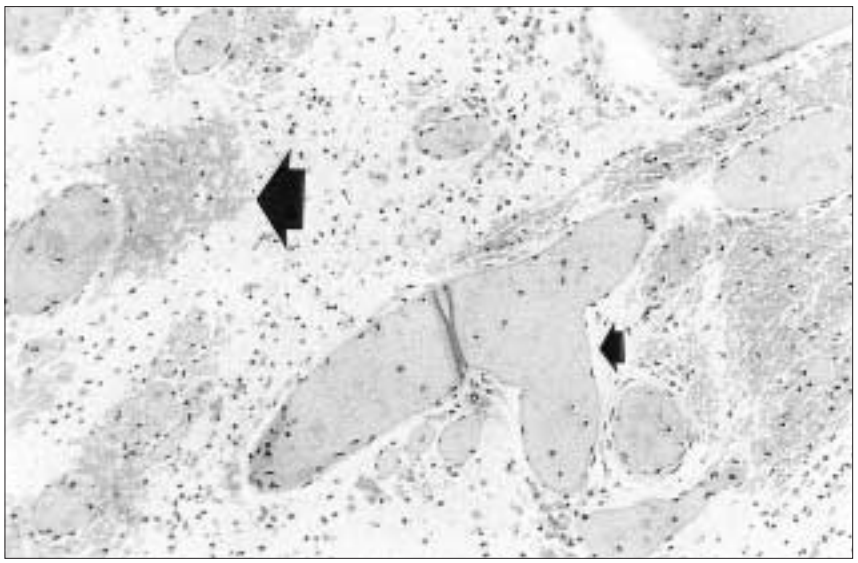

FIG. 2. Hemorrhage into the subcutaneous neurofibroma (large arrow). Note the thin-walled vasculature within the resected specimen (small arrow) (hematoxylin–eosin, original magnification × 25).

or vascular invasion by the neurofibroma. However, thin-walled vessels were observed in the neurofibroma adjacent to areas of hemorrhage (Fig. 2).

COMMENT

There is a well-documented tendency toward bleeding in plexiform neurofibromas in patients with von Recklinghausen's disease.[1,3] However, massive life-threatening hemorrhage is rare. There have been reports of fatal or near-fatal spontaneous hemothoraces in patients with NF-1,[3] but to our knowledge, an expanding subcutaneous chest-wall hematoma causing hemodynamic instability has not been reported.

Patients with NF-1 are at increased risk for severe bleeding from a spontaneous source or secondary to mild trauma. The perioperative course of these patients may be complicated by a significant bleeding diathesis. Our patient presented with a mild traumatic injury to a cutaneous neurofibroma, which rapidly progressed into a life-threatening hemorrhage. This patient was managed with aggressive surgical débridement and hemostasis with prompt correction of her underlying coagulopathy. Despite her prolonged hospital stay, we believe that this aggressive surgical approach contributed to her favourable outcome.

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