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Case report

Life-threatening intratumoral hemorrhage in plexiform neurofibroma: A case report

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ABSTRACT

Neurofibromatosis type 1 (von Recklinghausen's disease) is an inherited nervous system disease affecting around 1 in 3000 and is characterized by café-au-lait spots, pigmented hamartomas of the iris (Lisch nodules), and multiple neurofibromas. Plexiform neurofibromas can show intratumoral bleeding, and the fragile surrounding blood vessels and soft tissues carry a risk of escalation to lethal bleeding. We encountered a 59-year-old man with neurofibromatosis type 1 who presented with a rapidly growing massive hematoma in the left buttock after hitting it against a chair 12 hours earlier. He was tachycardic and hypotensive (hemoglobin, 5.7 g/dl), with severe swelling and tenderness in the left gluteal region. Plain computed tomography revealed a large, hyperdense, subcutaneous soft-tissue mass on the left buttock measuring 23 × 12 × 24 cm. Selective angiography demonstrated significant arterial supply to the lesion from a highly hypertrophied left L4 lumbar artery as well as the left superior gluteal artery. Embolization of these vessels was performed to reduce tumor blood supply. After returning to the intensive care unit, a central area of skin necrosis was noted. The wound was debrided; healing with scarring after 6 weeks with the tumor returning to its original size within 6 months. Hemorrhage in neurofibromatosis type 1 is attributed to a friable vasculature secondary to arterial dysplasia or vascular invasion by the neurofibroma. As neurofibroma is highly vascular, vessel rupture can occur spontaneously or with trivial trauma. Arterial embolization represents the method of choice for

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treating intratumoral bleeding in neurofibromatosis type 1, as a minimally invasive means of controlling arterial bleeding.

Introduction

Neurofibromatosis type 1 (NF1), also known as von Recklinghausen's disease, is an inherited disease of the nervous system with a frequency of around 1 in 3000.[1] This pathology shows three major features: café-au-lait spots; pigmented hamartomas of the iris (Lisch nodules); and multiple neurofibromas. The tendency toward bleeding within the plexiform neurofibromas is well recognized, but life-threatening hemorrhages are rare. Most reported cases have been associated with intrathoracic and gastrointestinal tumors,[2,3] but tumors of the face, chest wall, and sacral region have also been reported.[4–6]

Although hemostasis is necessary with intratumoral hemorrhage, surgical procedures are difficult due to the fragile nature of surrounding blood vessels and soft tissues and carry a risk of leading to lethal bleeding.[4,5,7,8]

We report the case of a patient with NF1 who experienced life-threatening intratumoral hemorrhage in the left buttock and was successfully treated with intravascular embolization.

Case report

A 59-year-old man with NF1 presented with a rapidly growing massive hematoma in the left buttock. Twelve hours prior to arrival, the patient felt pain in the region after he had hit his buttock against a chair. He was tachycardic and hypotensive, with a hemoglobin level of 5.7 g/dl. Severe swelling and tenderness were observed on the left buttock region (Figure 1). He received a transfusion of 2 liters of crystalloid fluid and 4 units of packed red blood cells and became hemodynamically stable with a blood pressure of 100/60 mmHg.

Plain computed tomography (CT) revealed a large, hyperdense, subcutaneous soft-tissue mass on the left buttock measuring 23 × 12 × 24 cm (Figure 2). As intratumoral bleeding was suspected, angiography was performed under local anesthesia via the right common femoral artery. Selective angiography demonstrated significant arterial supply to the lesion from a highly hypertrophied left L4 lumbar artery, as well as the left superior gluteal artery as a major branch of the left internal iliac artery (Figure 3). No extravasation of contrast material was identified, but all feeding vessels were tortuous

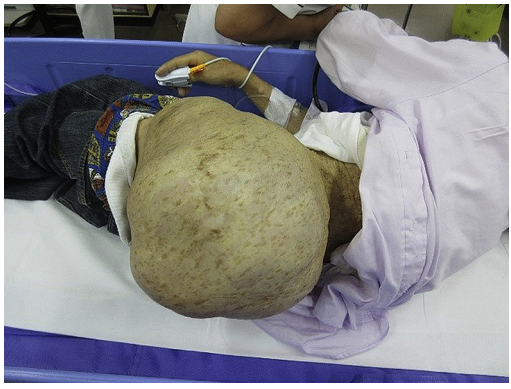

Figure 1. Appearance of the mass lesion on arrival.