Intravascular pyogenic granuloma in a child

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Intravascular pyogenic granuloma (IVPG) is a rare benign intraluminal vascular tumour. It was first described by Cooper et al in 1979 (1). Histologically, it appears identical to the significantly more common and well-known variety of pyogenic granuloma that occurs on epithelial-lined surfaces such as the skin and oral mucosa; however, the capillary proliferation is entirely intravascular in IVPG. Clinically, it most commonly presents in middle age as a nonspecific mass in the veins of the neck or upper limb (1). There are few case reports of this tumour in children.

CASE PRESENTATION

A 14-year-old male presented with an eight-month history of a painless lump on the radial aspect of his distal right forearm. There was no specific history of trauma. Ten months previously, he had had an enchondroma excised from his right middle finger. On clinical examination, there was a nontender mobile fusiform subcutaneous mass measuring approximately 10 mm × 5 mm. There were no skin changes overlying the lesion. However, there were faint café au lait macules noted on the patient’s neck and medial aspect of the right upper arm. Before presentation at The Hospital for Sick Children (Toronto, Ontario), an ultrasound of the lesion had been performed showing a solid lesion measuring 20 mm × 8 mm × 4 mm. The suggested differential diagnosis from the ultrasound included epidermal inclusion cyst, ganglion cyst or schwannoma. Considering the patient’s café au lait macules, preoperatively the mass was believed to most likely represent a nerve sheath tumour associated with the superficial radial nerve.

Under general anesthesia and tourniquet control, a longitudinal incision was made directly over the lesion. The tumour was found to be within the cephalic vein (Figure 1). A segment of the cephalic vein was resected including the tumour and a portion of macroscopically normal vein at each margin (Figure 2).

Histology revealed that the lesion within the vein contained a proliferation of capillary-size channels arranged in lobules separated by fibrous connective tissue. The lesion was attached to the vein wall at one aspect where there was loss of the normal muscle layer of the vein. The lesion was positive for CD31 and negative for GLUT1 and D2-40. This appearance is consistent with an IVPG.

Key Words: Intravascular pyogenic granuloma; Pediatric; Lobular capillary hemangioma

Intravascular pyogenic granuloma is a rare tumour, with approximately 30 cases presented previously in the literature. It most commonly occurs in the veins of the head and neck and upper extremity in middle-age patients. There is a paucity of reports detailing this lesion the pediatric literature. A case involving a 14-year-old boy with an intravenous pyogenic granuloma is presented.

DISCUSSION

Pyogenic granuloma is a common acquired benign vascular tumour (2). It classically occurs on epithelial lined surfaces such as skin and oral and genital mucosa. They are common in children. They present as a rapidly developing, sessile or pedunculated, red-purple mass prone to ulceration and bleeding (2).
In contrast, IVPG are rare intravascular tumours (1,3-5). Histologically, they are characterized by a lobular proliferation of capillaries similar in appearance to the more common cutaneous pyogenic granulomas (1). They differ from their cutaneous counterparts by being confined within the lumen of a vein and have a sparse inflammatory cell infiltrate. They most commonly occur within the veins of the head and neck and upper extremities.

Cooper et al (1) first described 18 cases of a previously unrecognized entity and coined the term intravenous pyogenic granuloma. Their study identified typical findings of intraluminal polyps attached to vein walls by fibrovascular stalks. These polyps were composed of lobules of capillaries separated by sparse spindle cells in a fibromyxoid stoma. This appearance is identical to uncomplicated pyogenic granulomas. IVPG lack a significant inflammatory infiltrate, which is a common secondary feature in mucocutaneous pyogenic granulomas that tend to be frequently traumatised.

The case presented in the present report demonstrates several similarities and differences compared with previously reported cases. Similar to this patient, IVPG have most commonly been reported in the upper extremity (1,6,7). They most commonly present as a non-specific asymptomatic subcutaneous mass. There is usually no history of previous trauma. There are no characteristic features of the presentation that allow a preoperative diagnosis to be made. Diagnosis relies on histopathological examination.

The patient presented in the present case report was considerably younger than most of the previously presented cases (3-7). In contrast to the more common mucocutaneous pyogenic granulomas, IVPG is unusual in children. Most of the reported cases occur in middle age. In the original study of IVPG, the mean age at presentation was 38 years (1).

Also, there have been no previous reports of IVPG in association with a history of enchondroma. Multiple enchondromas have been described in association with vascular malformations and café au lait macules in Mafucci syndrome. The case presented in the present report involved a single enchondroma and single vascular anomaly with café au lait macules. While this is clearly not a case of Mafucci syndrome and probably represents a coincidental occurrence of these lesions, this patient will continue to be followed.

The pathogenesis of IVPG is unknown (1,4). It is a benign lesion that demonstrates no tendency for hematogenous spread. Because the diagnosis of IVPG cannot be made clinically, the importance of IVPG lies in its histological differentiation from other intravascular lesions including vegetant intravascular hemangioendothelioma, inflammatory, angiomatous nodule and angiosarcoma (1,3-5). From a surgical perspective, excision of the lesion with a margin of macroscopically normal vein appears curative.

DISCLOSURES: None to report.

REFERENCES