Successful treatment with interferon alfa in infiltrating angiolipoma: a case presenting with Kasabach-Merritt syndrome

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We report a case of infiltrating angiolipoma who presented with a large mass on the shoulder and the signs of Kasabach-Merritt syndrome. She improved dramatically within one month and the mass completely disappeared after six months of interferon treatment.

Infiltrating angiolipoma is a rare benign tumour characterised by infiltration of skeletal muscle by adipose tissue combined with angiomatous elements. Infiltrating angiolipomas are usually unresectable surgically. Curative surgery is very mutilating in most cases. There is no well known alternative treatment for such unresectable infiltrating angiolipomas.

Interferons have been used therapeutically in a number of angiomatous diseases such as hemangiomas. A beneficial response to interferon therapy in life threatening hemangiomas has been reported. We present a case of infiltrating angiolipoma who presented with thrombocytopenic purpura; interferon alfa was used to shrink the large mass on the shoulder.

CASE REPORT

A 2 year old girl was admitted to Selçuk University Hospital with painful swelling on the right shoulder and several ecchymotic lesions on her body. On examination, there was a 4×5 cm soft tissue mass surrounding the right shoulder and axillary region. The mass was tender and caused significant limitation of arm movement. There were many various sized purpuric and ecchymotic lesions over the trunk and extremities.

Complete blood count revealed significant anaemia and thrombocytopenia (haemoglobin 77 g/l, white blood cells 7.6×10^9/l, platelets 27×10^9/l). Blood smear findings were normal except for the absence of platelets. Bone marrow examination showed normocellular with an increase in megakaryocytes and erythroid hyperplasia. The coagulation profile revealed prolonged prothrombin and partial thromboplastin time, decreased fibrinogen, and increased fibrin degradation products. (prothrombin time 14.1 seconds; partial thromboplastin time 46.3 seconds; fibrinogen 2.7 µmol/l; fibrin degradation products >2000).

High dose methylprednisolone was initiated with the presumptive diagnosis of acute idiopathic thrombocytopenic purpura. The platelet count gradually increased after the first week of steroid therapy. The steroid dose was tapered and discontinued within two weeks. One month later she represented with complaints of a progressively enlarging mass on her shoulder and ecchymotic lesions on her body. Magnetic resonance imaging (MRI) showed an ill defined infiltrating solid mass with high signal intensity involving the anterior part of the right scapula, extending into the proximal metaphysis of the right humerus (fig 1). On pathological examination of incisional biopsy, the mass was seen to be composed of mature adipose tissue, fibrous bands, skeletal muscle cells, and vascular tissue. She was referred to our hospital for further evaluation and treatment. Pathological findings led to a diagnosis of angiolipoma.

**Figure 1** Initial MRI of right shoulder, showing infiltrating solid mass involving the anterior part of the right scapula.

**Figure 2** Post-treatment MRI of right shoulder, showing shrinkage of the mass.
Since the mass was considered unresectable, high dose methylprednisolone was restarted to shrink the tumour. Although the size of the mass and thrombocytopenia improved dramatically within two months, all the symptoms reappeared when the steroid dose was reduced. Interferon alpha-2a (Roferon) 3 million units/m² three times a week was started. The platelet count increased gradually and the coagulation profile improved within one month. The mass reduced to 50% of its initial size within three months of treatment and completely disappeared after six months (fig 2). Interferon exhibited its therapeutic effect by inhibiting angiogenesis and endothelial cell migration and proliferation. It may also show beneficial effects in Kasabach-Merritt syndrome by decreasing platelet adherence and trapping by the endothelial cell surface. We used interferon in this case as the mass and its complications recurred after steroid tapering. Laboratory findings, including haemoglobin, platelet count, and coagulation profile resolved within one month. The volume of the mass also dramatically decreased to about 50% of the initial size within three months. To our knowledge, this is the first reported case of infiltrating angiolipoma presenting with Kasabach-Merritt syndrome in a child that responded to interferon therapy.

In conclusion, interferon may be considered in the treatment of various benign tumours containing vascular elements such as angiolipomas.

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REFERENCES
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