CASE REPORT

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Pigmented villonodular synovitis of the ankle in an adolescent

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Abstract We report a case of pigmented villonodular synovitis (PVNS) in an adolescent with monarticular involvement of the ankle and without congenital anomalies or sibling involvement.

Résumé Les auteurs rapportent un cas de synovite villonodulaire pigmentée de la cheville, chez une adolescente de 14 ans, ne présentant aucun facteur d'anomalie congénitale ni de facteur familial.

Introduction

Pigmented villonodular synovitis (PVNS) is a rare condition of unknown aetiology. It occurs most commonly in the knee but rarely in the ankle [12, 14]. Moreover, solitary PVNS is uncommon in children [1, 2, 3, 4, 13] where it is usually associated either with congenital anomalies or with sibling involvement [5, 6, 7, 8, 9]. There is a low rate of malignant change [4] but the lesion has the ability for reactivated growth and a tendency to recur [5]. This paper reports the case of a 14-year-old girl who presented with ankle swelling due to a solitary PVNS.

Case report

A healthy 14-year-old girl presented at our institution with a 1-year history of left ankle swelling and tenderness. Her symptoms had progressed during the previous 3 months with a history of 2 months of increasing pain and swelling in the left ankle although this had not limited her activities. The patient's past medical history and family history were unremarkable.

Examination revealed a mass measuring 4 cm×5 cm in the left ankle lying posterior to the lateral malleolus. The ankle moved normally and her neurovascular status was intact. A blood count, sedimentation rate, rheumatoid factor and coagulation time were within normal limits.

Diagnostic imaging included plain radiographs of the left ankle and these revealed a "subtle" soft tissue swelling inferior to the lateral malleolus. T1-weighted coronal (Fig. 1) and axial (Fig. 2) MRI showed a predominantly intermediate signal mass extending laterally from between the calcaneus and navicular. On the T-weighted axial image the mass had a more heterogeneous appearance with linear areas of high and very low signal. No bone erosions were seen in either the plain radiographs or the MRI.

An excisional biopsy was performed under tourniquet control through a transverse lateral ankle incision, and at surgery a 3 cm×4 cm nodular, red, brown and yellow pigmented mass was excised from the left ankle joint (Fig. 3). Both the tibial and talar joint surfaces were intact. The gross specimen was characteristic of PVNS, and this was confirmed by the histological findings of typical PVNS with the presence of foam cells, large multinucleated giant cells and iron deposits. Thus our diagnosis was confirmed by the histopathological findings and MRI.

The patient's symptoms improved significantly following surgery, but she was left with mild pain when walking. This was still present at the most recent clinical follow-up some 2 years after excision. Three years and 4 months after surgery MRI showed that the tumour, had completely disappeared.

Discussion

PVNS of the ankle is much rarer than that of the knee. It was found in the foot and ankle in approximately 2.5% of the cases of Rao et al. [12] and 25% of those of Ushijima [14]. The first reported case of PVNS of the ankle was

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T2-weighted coronal

Fig. 1 T2-weighted coronal MR images: relatively hyperintense mass

T1-weighted axial

Fig. 2 T1-weighted axial MR images: muscle isointense mass with some hypointense area

Fig. 3 Intraoperative findings during synovectomy. The synovial tissue of the ankle joint shows a thickened synovial layer with brownish and yellowish villonodular surface

Table 1 Pigmented villonodular synovitis in childhood

	No.	Patient	Ref. No.	Age	Gender	Location of lesion	Associated conditions
1968	1	Bobechko et al.	2	4	M	(L) Knee	intraarticular and cutaneous hemangiomas
	2	"		5	M	(R) Knee	intraarticular and cutaneous hemangiomas
	3	"		10	F	(R) Knee	intraarticular and cutaneous hemangiomas
1975	4	Leszczynski et.al.	7	7	F	(R) Knee (Bil) Ankle	pulmonary stenosis hemangioma of the lip splenomegaly
1977	5	Lindenbaum et.al.	8	14	F	(Bil) Knee (Bil) Ankle (Bil) Elbow	pulmonary abnormality scoliosis
1981	6	Wagner et.al.	16	9	F	(Bil) Knee (Bil) Ankle (Bil) Wrist	borderlin intelligence giant cell reparative granuloma of the mandible, scoliosis
1985	7	Walls et al.	17	6	F	(R) Knee (Bil)	fetal hydrantoin syndrome at birth, hypertelorism, a broad nasal bridge
1986	8	Wendt et al.	19	13	F	(Bil) Knee (R) Wrist (L) Foot	a high arched palate, pulmonary stenosis multiple lentigines syndrome, (congenital anomalies of the cardiac, genitourinary, endocrine, musculo skeletal and neurologic systems.)
	9	"		10	M	(R) Ankle (R) Wrist (L) Talus	multiple lentigines syndrome
1988	10	Aghasi et.al.	1	3	M	(L) Ankle	monarticular
1993	11	Soifer et.al.	13	11	F	(L) Ankle	monarticular
	12	Bruns et.al.	3	11	F	Knee	monarticular
1996	13	Kay et.al.	6	11	M	(L) Wrist (R) Knee	
1998	14	Minisola	9	13	F	multiple Posterior tibialis tendon	Noonanlike or multiple giant cell lesion syndrome, short stature, blue sclerae flatnened nose, low set ears, hypertelorism a short neck, curly hair, hyperkeratonic skin, hypoplastic genitalia, pulmonary sterosis congenital anomalies of the genitourinary tract
	15	Vedantan	15	4	M	(Bil) Knee (Bil) Ankle	
	16	Ganley	4	10	F	(R) Ankle	monarticular
2000	17	present case		14	F	(L) Ankle	monarticular

No., number of patient; Ref. No., number of reference

made in 1975 by Leszczynski [7]. Since then there have been a few hundred cases of PVNS of the ankle reported in the literature [4, 11] but in only 9 of 15 cases of PVNS occurring in children (Table 1). PVNS usually affects adults during their third or fourth decade in life [10]. The

reported cases of PVNS occurring in childhood (Table 1) consist of 5 boys and 10 girls whose ages ranged from 3 to 14 years at the time of presentation. In cases occurring in childhood PVNS is usually associated either with congenital anomalies or sibling involvement [3, 5].

Physical examination of a patient with PVNS may demonstrate swelling, tenderness, erythema, a limited range of motion or even a palpable mass. Aspiration of an affected joint may reveal serous fluid or even frank blood although this is rare in the ankle.

MRI may be the most effective means of detecting PVNS and this is due to the presence of a haemosiderin which causes a dose-related decrease in the relaxation time of T1 and T2 images. A sufficient level of haemosiderin will cause T1 and T2 images to appear dark [4, 15]. To confirm the diagnosis either a needle or an open biopsy (often excisional) is necessary.

Histological examination of the specimen reveals villous and/or nodular proliferations of the synovium. In the diffuse form there are villous synovial fronds infiltrated and filled by the proliferating fibro-histiocytic and giant cell reaction. The localised form consists of focal aggregates of nodules.

Surgical treatment should be performed as soon as possible in order to prevent cartilage infiltration which can occur [11, 16].

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