J Hong Kong Col Radiol. 2010;13:202-4

CASE REPORT

Role of Magnetic Resonance Imaging in Popliteal Pterygium Syndrome

D Chinchure[1], A Littooij[2]

[1]Department of Diagnostic Radiology, Khoo Teck Puat Hospital; [2]Department of Diagnostic Imaging, KK Women's and Children's Hospital, Singapore

ABSTRACT

We describe magnetic resonance imaging in the preoperative evaluation of children with popliteal pterygium syndrome to identify the position of the popliteal artery and peroneal nerve. A nine-year-old boy, with known popliteal pterygium syndrome, had a problem straightening both knees since birth. This was due to bands connecting his hamstrings to his calf muscles. He was referred for magnetic resonance imaging to evaluate the position of popliteal arteries and peroneal nerves in relation to the soft tissue bands. The magnetic resonance imaging delineated the position of the popliteal artery and peroneal nerve accurately with respect to the pterygium band. This information is crucial in the preoperative planning of treatment for children with popliteal pterygium syndrome.

Key Words: *Magnetic resonance imaging; Peroneal nerve; Popliteal artery; Pterygium*

中文摘要

磁共振成像在診斷腘翼狀胬肉綜合徵中的作用

D Chinchure、A Littooij

本文報告一宗利用磁共振成像為患有腘翼狀胬肉綜合徵的兒童進行術前評估以確定腘動脈及腓總神經位置的病例。一名9歲男童患有腘翼狀胬肉綜合徵，由於他的大腿後側肌群及小腿後側肌群有帶狀連接，自出生後兩膝關節都不能伸展。病人經轉介接受磁共振成像檢查，以評估腘動脈及腓總神經的位置及與軟組織帶的關係。磁共振成像能準確地勾畫出相對於翼狀贅片，腘動脈及腓總神經的位置。這些資料對腘翼狀胬肉綜合徵兒童患者進行術前評估相當重要。

INTRODUCTION

Popliteal pterygium syndrome is a congenital anomaly in which a connective tissue band extends from the ischium to calcaneum causing flexion deformity of the knee joint, and is associated with genitourinary and craniofacial anomalies. Treatment entails surgical resection of the pterygium to relieve the flexion deformity of the knee joint.[1-4] In the preoperative evaluation, knowledge as to the positions of vessels and nerves in relation to the connective tissue band is crucial, in order to avoid damaging these structures. In these patients, the popliteal artery and peroneal nerve may be normally placed or have an abnormal course closely related to the connective tissue band.[1] The peroneal nerve is most likely to be involved by the pterygium band. We describe a patient having a popliteal pterygium syndrome, where magnetic resonance imaging (MRI) was used to evaluate the

Correspondence: *Dr Dinesh Chinchure, Department of Diagnostic Radiology, Khoo Teck Puat Hospital, Singapore. Tel: (65) 8126 3447; Fax: (65) 6602 3796; Email: drdineshdc@yahoo.com*

Submitted: 6 Sep 2010; Accepted: 13 Sep 2010.

position of popliteal artery and peroneal nerve.

CASE REPORT

A nine-year-old boy was referred for MRI of both lower limbs in June 2010 to evaluate the flexion deformity of both knees since birth. A subcutaneous soft tissue band connecting the hamstrings to the calf muscles was noted. Aged one year, he had undergone skin distraction surgery of both knees. Initial radiographs revealed flexion deformity of both knees with absent patellae (Figure 1). MRI of both lower limbs was performed using 1.5-Tesla GE HDxT Excite scanner (General Electric Medical Systems, Milwaukee, WI, USA). A torso-phased array coil was used to include pelvis to foot in the field of view. Multiple sequences were obtained including post-contrast images. The extent and signal characteristics of the soft tissue bands and positions of both popliteal arteries and peroneal nerves in relation to each other were studied. The pterygia were noted on the MRI as posterior subcutaneous soft tissue bands arising from the ischium and terminating on the calcaneum, after passing through the posterior aspect of the popliteal fossa. The signal characteristics of the pterygia corresponded to fibrous tissue, with its thickest part in the popliteal fossa. The muscles were atrophic due to chronic disuse. In both lower limbs, the popliteal artery was in a normal position deep to the pterygium band and adequate cleavage planes were evident (Figure 2). The positions of peroneal nerves were also assessed. The left peroneal nerve was involved, and it was seen to be abutting the anterior margin of pterygium (Figure 3). The right peroneal nerve and both posterior tibial nerves were not involved with the band of pterygium (images not shown).

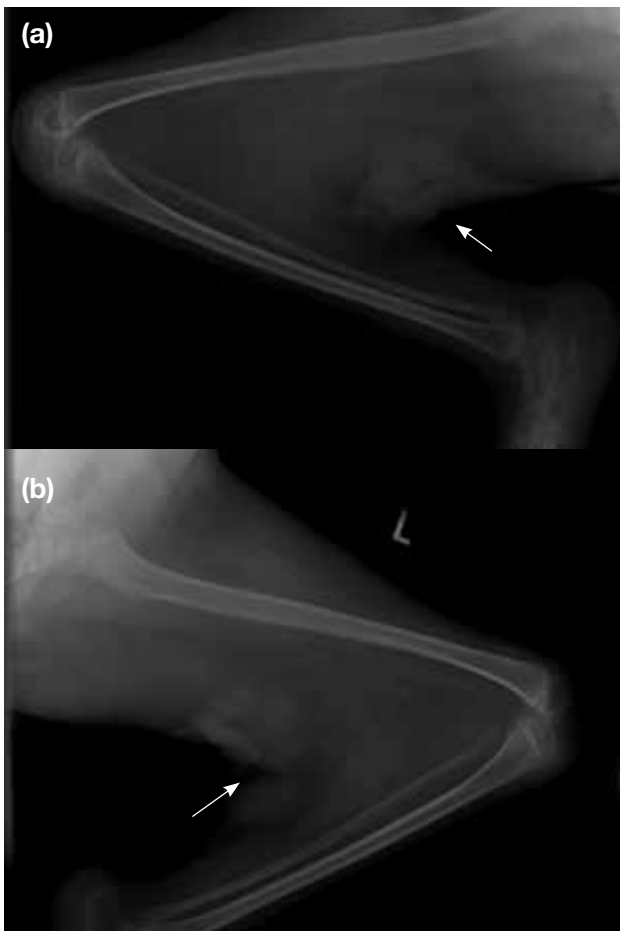

Figure 1. Lateral radiographs of (a) right and (b) left knees showing marked flexion deformity and absent patellae. Note that the pterygium bands are seen as soft tissue opacities along the subcutaneous planes of both popliteal fossae (arrows).

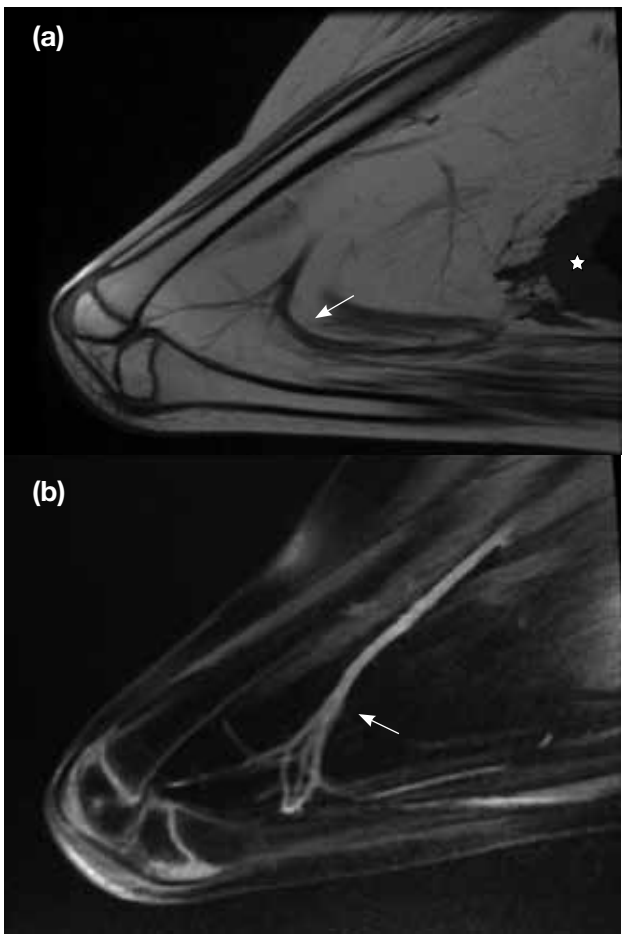

Figure 2. (a) A sagittal T1-weighted magnetic resonance image (MRI) of the left knee, and (b) post-contrast fat-suppressed sagittal T1-weighted MRI of the right knee. These show normal positions of the popliteal arteries (arrows) away from the thickened subcutaneous soft tissue pterygium bands (*).

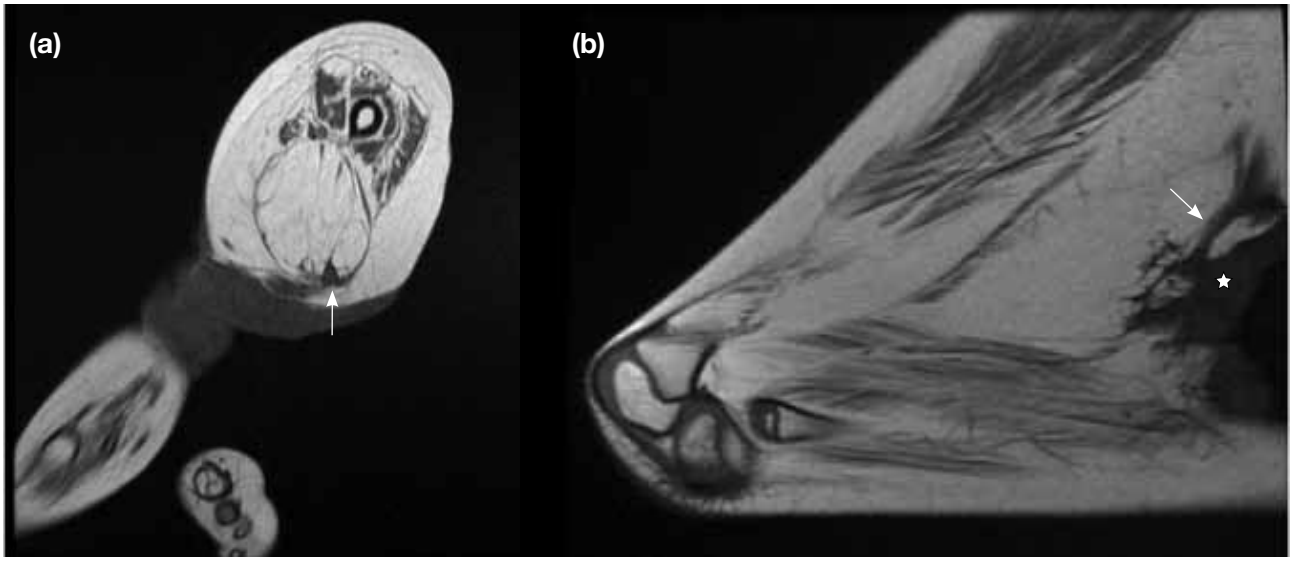

Figure 3. (a) Axial and (b) sagittal T1-weighted magnetic resonance images of the left knee showing the abnormal position of the peroneal nerve (arrows) abutting the anterior margin of pterygium band (*).

DISCUSSION

The word 'pterygium' is derived from the Greek word *pterygion*, which means wing. Pathologically it denotes a wing-like abnormal band of tissue. The common locations for pterygia are the neck, the eye, the digits, and joints (such as the knee and elbow).[2] The neck pterygium or webbed neck is associated with trisomy 21, 13 and Turner's syndrome. Escobar syndrome is associated with multiple pterygia. The popliteal pterygium syndrome is a rare condition, in which the patient has facial, genitourinary and skeletal anomalies along with popliteal pterygium. It usually shows autosomal dominant inheritance. The soft tissue band associated with this syndrome can be complete or incomplete, and extends from the ischium to the calcaneum.[1] When bilateral, their thickness and extent can be symmetrical or asymmetrical. Typically they cause flexion deformity at the knees limiting their movement. Surgical resection of these bands is essential to relieve the contracture. The key structures including popliteal arteries and personal nerves can be abnormally and lie within or just adjacent to the pterygium.[1-4] Precise anatomical localisation of these structures is essential before surgical resection is planned, so as to avoid damaging them during surgery. Doppler ultrasound and computed tomography may be used to delineate the anatomy. The former is limited by operator dependence, particularly due to difficulty posed by the contracted knees. Computed tomography accurately identifies the popliteal artery, but peroneal nerve identification may be more difficult. MRI therefore plays a crucial role in presurgical evaluation of patients with popliteal pterygium syndrome, due to its excellent soft tissue resolution, especially with respect to nerves.[5] Moreover, it is a preferred tool as it does not involve ionising radiation, which is of particular importance to these patients as they are in the paediatric age-group.

CONCLUSION

MRI is the modality of choice in the presurgical evaluation of children with popliteal pterygium syndrome, as it accurately delineates the position of popliteal artery and peroneal nerve with respect to the pterygium.

REFERENCES

1. Oppenheim WL, Larson KR, McNabb MB, Smith CF, Setoguchi Y. Popliteal pterygium syndrome: an orthopaedic perspective. J Pediatr Orthop. 1990;10:58-64.
2. Taybi H, Lachman RS. Radiology of syndromes, metabolic disorders and skeletal dysplasias. 4th ed. St. Louis: Mosby; 1996: 411-3.
3. Froster-Iskenius UG. Popliteal pterygium syndrome. J Med Genet. 1990;27:320-6.
4. Maral T, Tuncali D, Ozgür F, Safak T, Gürsu KG. A case of popliteal pterygium treated along with nerve expansion. Plast Reconstr Surg. 1997;100:91-5.
5. Donnelly LF, Emery KH, Do TT. MR imaging of popliteal pterygium syndrome in pediatric patients. AJR Am J Roentgenol. 2002;178:1281-4.