age of diagnosis was 5 years with a male predominance (76%), the majority with unilateral disease (95%). Aggressive management for early cases, including laser photocoagulation, cryotherapy, and drainage of subretinal exudates using pars plana vitrectomy techniques, may prevent neovascular glaucoma and a painful blind eye.

Because the definitive genetic defects for both VATER association and Coats disease in humans are unknown, we can only speculate about the possible common genetic link. Recently, an adriamycin-induced rat model of the VATER association, with defects in the hedgehog gene pathway, has been developed. The secreted glycoprotein, sonic hedgehog (SHH), acts as an endodermal signal that controls gut and lung patterning. Interestingly, the SHH protein is also secreted by retinal ganglion cells to help optic disc astrocyte precursor cells to guide retinal axon growth, and to convert optic stalk neuroepithelial cells into pigment cells. Both VATER and retinal defects can be produced by targeted SHH mutations. Ultrastructural retinal cell abnormalities are not infrequently found in pathologically examined eyes of patients with Coats disease. Nevertheless, it remains to be elucidated whether any aberrations in these pathways may explain the unusual clustering of specific defects in our case.

Charmaine Hon, FRCS
Tak-chuen Ko, FRCS

The authors have no relevant financial interest in this article.

Correspondence: Dr Hon, Department of Ophthalmology, Prince of Wales Hospital, Shatin, Hong Kong (honz@ha.org.hk).


Idiopathic Isolated Abscess in an Extraocular Muscle in a Child

An idiopathic muscle abscess is a rare condition that should be considered in the differential diagnosis of painful limitations of eye movements and double vision in children.

We herein present the case of an idiopathic muscle abscess in an 11-year-old girl. Myositis was initially suspected, but the magnetic resonance imaging (MRI) scan showed typical signs of an abscess. Antibiotic therapy led to a rapid and complete improvement of all symptoms. To our knowledge, this is the first report of an idiopathic isolated abscess in an extraocular muscle in a child.

Figure 1. Patient photographs before (A) and after (B) treatment.

Figure 2. Oblique sagittal (A) and axial (B) T1-weighted contrast-enhanced spin-echo magnetic resonance imaging (MRI) scans. Diffuse swelling of the left superior rectus muscle is seen surrounding an abscess (diameter, 8 mm), with central cavitation and rim of contrast enhancement of about 3 mm surrounded by a diffuse swelling of the left superior rectus muscle (Figure 2A and B).
The measured serological variables (including adenovirus, human herpesvirus 6, herpes simplex virus, varicella-zoster virus, enterovirus, Borrelia burgdorferi, Epstein-Barr virus, measles virus, human immunodeficiency virus, syphilis, and cysticercosis) and cerebrospinal fluid variables (including sediment, glucose and protein levels, and serological findings for B burgdorferi and Russian spring-summer encephalitis) were within reference range.

Immediately after the first visit, high-dose intravenous antibiotic therapy was introduced (combination of amoxicillin sodium and potassium clavulanate, 8.8 g/d for 10 days). No amelioration of symptoms could be observed after 2 days, so metronidazole (1.35 g/d for 5 days) and ceftriaxone disodium (2 g/d for 19 days) were added. This led to a rapid and complete improvement of all symptoms (visual acuity of 20/20 OU, free motility in all directions in both eyes without double vision, and no inflammatory signs, including eyelid edema [Figure 1B]). An MRI control scan, obtained 2 weeks after initiation of therapy, showed only a discrete residual swelling of the affected muscle (Figure 2C and D).

Comment. Main differential diagnoses in children are other inflammations and infections (eg, diffuse orbital pseudotumor, orbital cellulitis),\textsuperscript{1} structural lesions (eg, dermoid cyst),\textsuperscript{2} vascular neoplastic lesions (eg, capillary hemangioma, lymphangioma),\textsuperscript{3} lymphoproliferative diseases (eg, lymphocytic granuloma),\textsuperscript{4} neurogenic tumors (eg, neuroblastoma, plexiform neurofibroma),\textsuperscript{5} mesenchymal tumors (eg, rhabdomyosarcoma), and metastatic carcinoma.

Although the clinical features are frequently suggestive of myositis, they are nonspecific, and noninvasive investigations such as orbital ultrasonography and MRI scans are required for precise anatomical tissue localization and diagnosis. The role of ocular muscle biopsy is probably limited to atypical cases, or those unresponsive to therapy, particularly to exclude neoplasia.

Timo Haufschild, MD
Peter Weber, MD
Ines Nuttli, MD
Burkhard Hecker, MD
Josef Flammer, MD
Hedwig J. Kaiser, MD

The authors have no relevant financial interest in this article.

Correspondence: Dr Flammer, University Eye Clinic, Mittlere Strasse 91, PO Box, CH-4012 Basel, Switzerland (Josef.Flammer@uhbs.ch). Dr Haufschild may be contacted at THaufschild@uhbs.ch