Pathologic Study of Supernumerary Orbital Band in Type I Duane Syndrome

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Established Facts

- Accessory orbital bands are a rare finding and have been reported to produce strabismus.
- Most of the bands are labeled as fibrous or muscular on the basis of radiological evidence.

Novel Insights

- Accessory orbital bands that are predominantly fibrous seem to produce strabismus.
- Compressed collagen may appear red using Masson trichrome staining. In such cases, identification may require polarization, Sirius red staining, and immunohistochemistry.

Keywords

Supernumerary band · Accessory orbital band · Histopathology · Strabismus · Duane syndrome

Abstract

Background/Aims: Accessory orbital bands are relatively rare and very few reports detail histopathology. Cases in the literature describe the composition of the bands as muscular and/or fibrous. The composition of the supernumerary band lying deep in the medial rectus muscle in a patient with type I Duane syndrome was investigated. **Methods:** Histochemical stains were used in conjunction with polarized light for differ-

entiating compressed collagen from muscle. Immunohistochemistry was used for verification of the presence of muscle.

Results: Compressed collagen appeared red using Masson trichrome staining. Collagen was positively identified by illumination with polarized light on several stains including the underutilized Sirius red dye. **Conclusions:** The findings of dense collagen fibers in the fibrotic band with focal striated muscle correlated with the restrictive strabismus. In concert with other cases in the literature, it is proposed that the fibrous bands are generally associated with restrictive strabismus. Bands that are muscular may or may not be associated with strabismus. Special techniques are needed to positively identify compressed collagen.

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Introduction

Accessory bands are rare orbital structures associated with restrictive strabismus. The bands are usually described as an incidental intraoperative finding or with the use of high-resolution orbital imaging [1]. Very few reports have described detailed histopathology of these supernumerary bands. The histopathology may provide insight into possible mechanisms for strabismus in such cases [2, 3]. Fibrosis following injury is preceded or accompanied by inflammatory infiltrates. We here present a case of restrictive strabismus due to an accessory band in Duane syndrome that underwent detailed histopathological examination. The findings have implications for the approach to strabismus in these patients.

Case Report

The subject was a 35-year-old man with symptomatic diplopia and strabismus. He had previously been diagnosed with esotropia (ET) secondary to type I left Duane syndrome and had no history of previous ocular surgery or trauma. His past medical, family and social histories were noncontributory.

On ophthalmic examination, best corrected visual acuity was 20/20 and 20/50 in the right and left eye, respectively. Stereopsis measured 40 s of arc at near with a significant left head turn. A marked limitation to abduction with narrowing of the palpebral fissure was observed in the left eye. Full ocular motility was observed in the right eye. Prisms demonstrated 20Δ of left (ET) and 2Δ of left hypotropia in the primary position, 6Δ of left ET in up gaze and 20Δ of left ET in down gaze. ET significantly increased in left gaze. He was orthotropic in right gaze. Slit lamp and dilated fundus examinations were unremarkable in both eyes.

The patient underwent preoperative forced duction testing. Severe restriction to abduction was apparent in the left eye. A paralimbal conjunctival incision was made. An extremely tight medial rectus muscle was found inserted 5.5 mm posterior to the limbus. A spring-loaded muscle clamp was placed on the medial rectus muscle and the muscle was disinserted from the sclera. A 6-0 polyglactin 910 suture was secured on the margin of the medial rectus muscle. Prior to reinserting the muscle, forced duction testing was repeated after removal of the eyelid speculum. Severe restriction to abduction persisted, despite complete disinsertion of the medial rectus muscle. Careful inspection of the sclera revealed an accessory band, distinct from the retracted muscle (Fig. 1). The band maintained a separate insertion, approximately 2 mm posterior to the muscle insertion and extended deep into the orbit. Following detachment and excision of this band, the forced duction test showed no remaining restriction in abduction. The left medial rectus muscle was recessed 4.5 mm posterior to its insertion. Attention then shifted to the right eye. The right medial rectus muscle was recessed 5.5 mm and the lateral rectus muscle was recessed 6.5 mm with an adjustable suture technique.

Histopathologic Studies

The accessory band was fixed in formalin and processed for paraffin embedment. Step sections were taken for histopathologic study including hematoxylin and eosin, Masson trichrome stain, elastic Masson trichrome, and Sirius red stains as previously described [4–6]. Polarizing microscopy was performed on stained sections to further distinguish collagen from smooth muscle. Immunohistochemistry for reactivity to anti-muscle-specific actin was used to verify the presence or absence of muscle. Appropriate tissues served as positive and negative controls. In addition, deletion of the primary antibody served as an additional negative control.

Results

Sections stained with hematoxylin and eosin of the accessory band revealed closely packed fibers that showed birefringence on polarization. In one focal area, striated muscle was seen with faint cross-striations and peripheral nuclei (Fig. 2). However, most of the tissue appeared to be compressed fibrous tissue. The Masson trichrome stain showed marked red staining but also birefringence with polarized light (Fig. 3a, b). To clarify this apparent disparity, Sirius red staining was performed (Fig. 4a). Staining was markedly enhanced in most areas where the Masson trichrome appeared red. These areas demonstrated birefringence and dichroism with polarized light further supporting the presence of collagen (Fig. 4b). Immunohistochemistry for muscle-specific actin was performed and showed only small areas of reactivity in the section (Fig. 5).

Discussion/Conclusion

The key findings of this study are the evidence that supernumerary bands that induce strabismus are composed mainly of fibrous tissue. Further histological studies of fibrous tissue that are under tension should not rely solely on Masson trichrome as there may be a false-positive red staining for muscle.

The term “supernumerary band” is largely applied to most accessory, soft tissue structures in the orbit. The clinically published cases of supernumerary bands without histopathology mainly describe the atypical *muscular* bands; these are most commonly identified in the superno-nasal orbit [7–10]. However, our study shows mainly fibrous tissue in the band with exiguous striated and smooth muscle. Only 6 prior studies detail histopathology (Table 1). In a cadaver study from an apparently asymptomatic individual with accessory bands, the bands

Fig. 1. Perioperative finding of an accessory orbital band.

Fig. 2. Photomicrograph of the focal striated muscle in the resected band specimen. Hematoxylin and eosin.

Fig. 3. a Photomicrograph of a section shows predominant red staining fibers with the Masson trichrome stain. **b** Polarization of the trichrome-stained sections highlights an irregular fibrous architecture with birefringence in areas staining red.

were described as muscle slips fusing the superior and inferior rectus muscles. Histological examination showed a predominance of striated muscle fibers that were well organized [11]. Similarly, another study reported bilateral accessory muscles in a patient with Gorlin syndrome with large-angle exotropia [12]. In one of the bands, histopathology presumably showed striated muscle (images not provided in the paper). This band was described as

taut. Resection of this band, without resecting the contralateral band, resulted in release and in correction of the exotropia. The remaining 3 cases with histopathology suggest findings similar to our own. There was predominant fibrosis with only fragments of atrophied muscle in some instances [2, 3, 13].

To understand the composition of the supernumerary bands, it is imperative to distinguish fibrous tissue

Fig. 4. Photomicrograph of a section stained with Sirius red. **a** The sample shows both red and poorly stained regions. **b** Polarized light reveals the birefringence of the red-stained areas as well as dichroism in less well-stained areas.

from muscle. One case in the literature presumed the bands were fibrous on the basis of the hematoxylin and eosin stain [2]. Others have used a trichrome stain to distinguish collagen from muscle although the images were not provided [12, 13]. Clinically appearing muscular bands were proven to be fibrous tissue in at least one case [3]. The other cases with strabismus are variably described as fibrous and muscular, but histopathology was not performed (Table 1). The dense collagen in our study yielded a marked false-positive reaction that stained fibrous tissue red and might have been misinterpreted as muscle. The basis for the false red color in tri-

Fig. 5. Photomicrograph of immunohistochemistry with antibody to muscle-specific antigen shows only focal reactive areas. Most regions that correspond to red staining with the trichrome stain are negative for reactivity. The areas of reactivity matched the areas of striated muscle depicted on hematoxylin and eosin.

chrome staining lies in differential removal of red and blue dyes by polyacids based on the size of the stain molecules. Because collagen is porous, the red stain (Biebrich scarlet-acid fuchsin) is removed by polyacids and replaced by aniline blue. Muscle tissue is not well penetrated by the polyacids and remains red. However, in our case, the collagen stained red because the red dye was not displaced. We attribute this to the compressed nature of the fibrous tissue in the band that also accounted for its tough clinical appearance. Ocular pathologists frequently examine specimens for presumed slipped muscle and need a reliable stain to differentiate dense fibrous tissue from muscle. Sirius red is an excellent alternative histochemical stain that has excellent inter-observer reliability and reproducibility in renal specimens [4–6]. In our study, polarization-induced birefringence in both the hematoxylin and eosin and Masson trichrome stain alerted us that red areas may actually be collagen. This was confirmed in Sirius red stains. Polarization microscopy also enhanced the birefringent substructure of collagen in the Sirius red stain and showed dichroism in areas that poorly stained for Sirius red.

The clinical counterpart of the compressed collagen fibers in the accessory band may be the severe restrictive strabismus and progressive tightness of the medial rectus muscle. The key to diagnosis was the intraoperative examination and repeat forced duction testing. Prior to the ini-

Table 1. Details of previous reports on various accessory orbital bands

Author	Year	Diagnosis/ presentation	Per-operative findings	Histopathology technique	Histopathological findings
1 Lueder et al. [3]	1998	Vertical strabismus (hypotropia)	Pink, striated, 2 mm wide, muscular band 18 mm from limbus in inferomedial quadrant	H&E stain	Paucicellular strip of pale fibrous tissue with small fascicles of mature skeletal muscle
2 Spierer et al. [13]	1991	Exotropia	A fibrotic band 2.5 mm wide inserting onto sclera 1 mm from limbus extending posteriorly parallel and separate from lateral rectus muscle.	Masson trichrome and periodic acid Schiff stain	Fibrous tissue, partially dense and partially loose, with sections of capillaries and peripheral nerves. No muscle tissue was noted on Masson trichrome and PAS stains
3 Özkan et al. [2]	2007	Prosis and hypotropia	Fibrotic superior oblique and superior rectus muscles with an accessory tissue band passing toward the limbus	H&E stain	Band tissue and superior rectus muscle revealed fibrosis and degenerated collagenized connective tissue
4 Haladaj et al. [11]	2018	Cadaveric dissection of orbit	Accessory muscle band between superior and inferior rectus	H&E stain	Striated skeletal muscle
5 Dobbs et al. [12]	2011	Exotropia	Accessory muscle from annulus of Zinn inserting on the posterior eye	H&E stains not shown	Striated muscle (not shown)
6 Fleischer [17]	1907	Cadaveric dissection of orbit	Accessory muscle band	H&E stain	Striated muscle
7 Goldstein [14]	1964	Esotropia and hypotropia	Fibrous bands inferonasally and inferotemporally	None	None
8 McNeer et al. [8]	1965	Double elevator palsy	Anomalous slip of inferior rectus muscle	None	None
9 Prakash et al. [15]	1985	Hypotropia and exotropia	Fibrotic, cord-like, fused superior oblique and superior rectus that could not be separated	None	None
10 Valmaggia et al. [18]	1996	Elevation deficit	CT and MRI showed accessory, fusiform, well-defined, solid structure in the retrobulbar space that originated from the annulus of Zinn and traversed anteriorly to insert on the 9 posterior parts of the globe inferolateral to the optic nerve. Band was isointense as extraocular muscles on MRI	None	None
11 von Lüdinghausen [19]	1998	Cadaveric dissection	Bilateral anomalous supernumerary muscle bands originating from common tendinous ring and inserting into inferior rectus anteriorly with a muscle bridge connecting the band to superior rectus	None	None
12 Savino et al. [20]	1998	Absent sursumduction of the affected eye	Radiological studies (B-scan, CT, MRI) showed a muscular band originating from Zinn's ring, traversing intraconally and inserting onto sclera above inferior rectus	None	None
13 Baldeschi et al. [21]	2007	Graves' orbitopathy	Supernumerary extraocular muscle inserting onto sclera 5 mm laterally to the insertion of the inferior rectus muscle 7 mm from corneal limbus	None	None
14 Man et al. [22]	2009	Unilateral vertical retraction	MRI scan showed linear band from optical apex to posterior lower lateral sclera	None	None
15 Murthy [16]	2007	Unilateral fibrosis, blepharoptosis, and enophthalmos syndrome	Fibrous, soft tissue band 12 mm in length below the optic nerve extending from the globe to the orbital apex.	None	None
16 Molinari et al. [23]	2016	Duane syndrome	Anomalous orbital structures in patients with Duane syndrome that were labelled as an accessory extraocular muscle inserting near the optic nerve	None	None
17 Kighthlinger et al. [24]	2017	7 cases with different diagnoses	Radiological evidence of orbital bands that bridged the superior and inferior rectus muscles	None	None

tial conjunctival incision, the restricted eye had a positive forced duction test. After disinsertion of the medial rectus muscle, the tough band was clearly visualized and repeat forced duction test was again positive. It was not until the accessory band was disinserted that the eye moved freely. The clinical findings match the pathologic findings of predominant fibrous tissue and analysis of the special stains reveal some challenges with interpretation of stains.

Khitri and Demer [1] imaged accessory bands using high-resolution surface coil magnetic resonance imaging. They reported two cases with type III Duane syndrome that had bands superonasally along the levator complex. Because the bands were inaccessible deep in the orbit, they were not amenable to excision and histopathology. Some reports suggest the presence of accessory *fibrous* tissue bands but do not provide detailed histopathology [14, 15].

The histopathology of the cases of accessory bands may provide some mechanistic insight. For those patients with histopathology, the history suggests a congenital onset of strabismus. These patients invariably show no evidence inflammation of the muscle (Table 1). The fibrosis does not appear to result from a focal insult but rather a disorder of mesodermal development. Early diagnosis of accessory orbital bands is clinically challenging and sometimes not achievable; however, in patients with atypical strabismus, it is paramount to broaden the differential diagnosis to include these structures. Orbital imaging can considerably facilitate preoperative identification and allow for timely surgical planning [1, 13, 16]. Another telltale finding of supernumerary bands is the “traction-indentation” sign. When attempting to rotate the eye against restriction, the atypical band may produce significant opposition, causing localized pressure with secondary scleral indentation [2]. However, in patients with atypical restrictive strabismus and a supernumerary band, disinsertion of both the accessory tissue and weakening of the associated extraocular muscle is often required.

In conclusion, we emphasize the importance of careful peroperative examination and forced duction testing in cases of restrictive strabismus. A positive forced duction could possibly be caused by an accessory orbital band which should be excised to relieve the restriction. Our review indicates that to date the pathologically verified cases of fibrotic supernumerary bands had all been associated with strabismus, most characteristically hypotropia. However, strabismus may be caused by either muscular or fibrous bands. Histopathological studies with appropriate stains for collagen and the judicious use of polarization microscopy are helpful to elicit the composition of these accessory bands. The clinical history in concert with the histopathology suggest fibrosis of supernumerary bands occurring at birth implying a developmental rather than an acquired origin.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

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Author Contributions

M.H.A.: Manuscript writing, final critical review.

S.L.P.: Data collection, final critical review.

F.G.V.: Data collection, final critical review.

A.K.T.: Data collection, final critical review.

B.J.G.: Manuscript writing, data analysis, final critical review.

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