

ENLARGED MUSCLE STRABISMUS

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ABSTRACT

We present four young patients with unusual clinical patterns of strabismus and CT appearance suggestive of thyroid eye disease. The young age of these patients and the absence of any other clinical features of thyroid eye disease or thyroid disease do not really allow thyroid eye disease to be a credible explanation for the clinical and radiological findings. Muscle biopsy shows increased fibrous tissue, not typical of thyroid eye disease. We propose that this is a new clinical syndrome.

KEYWORDS

Strabismus, thyroid eye disease.

INTRODUCTION

We herein report a case series of four young patients with the following clinical features:

1. Unusual clinical patterns of strabismus.
2. Extraocular muscle enlargement on CT scanning.
3. No evidence in three of thyroid eye disease or any other conditions known to cause extraocular muscle enlargement. One patient showed minimal abnormalities of thyroid biochemistry.
4. Muscle biopsy in 2 patients showed focal increase in endomysial fibrous tissue when compared with age matched controls.

CASE 1

An 18 year old man presented with a 5 years history of constant horizontal diplopia. He was given glasses and orthoptic exercises at ages 6 and again at 14. With + 1.75 DS OU, the acuities were 6/6, N 3. He had a 15° esotropia. He had minimal vertical misalignments. He had LIO + 1, LSO - 1. There was no fundus torsion. With a prism in place he had 20 sec stereo. A non-contrast coronal CT showed bilateral symmetrical extraocular muscle enlargement in the posterior orbits. Thyroid biochemistry and immunology was normal. He was prism adapted to 30° ET and BMR of 4.5 mm OU was carried out.

A biopsy was taken 10 mm from the insertion. This showed a focal increase in endomysial fibrous tissue. EM showed lipofuscin in muscle fibres. Both of these findings are inappropriate for this age, lipofuscin being typically seen in patients over 30 and fibrous tissue in patients over 60.

Following the above surgery he developed a recurrence of his esotropia requiring prism.

CASE 2

This 27 year old male presented with diplopia on right gaze since childhood. He had a left hypertropia for distance greater on right gaze and greater in direction of action of LSO.

Left hypertropia for distance:

| | | | |
|------------|----|----|-------------|
| Up right | 18 | 12 | Up left |
| Right gaze | 30 | 20 | 8 Left gaze |
| Down right | 35 | 5 | Down left |

He had 10 degrees of fundus exocyclotorsion. He had diplopia in all directions of gaze to the right of the midline, and 10 degrees to the left of midline. He had left inferior oblique over action, and left superior oblique under action. On left gaze, he had a duction deficit of RIO, and of LIR. Non-contrast coronal CT scan shows bilateral symmetrical enlargement of the recti. The superior obliques are normal. Thyroid chemistry and immunology was normal. He has not yet had treatment for this diplopia.

CASE 3

This 6 year old female has had anomolous vertical movement of the right eye with a chin up head posture and retraction of the right lower lid since the first year of life. Forced duction test shows a tight RIR. She has an incomitant horizontal deviation with ET 10° on right gaze and XT 6° on left gaze. Coronal CT shows enlargement of the medial and inferior recti on both sides. Thyroid chemistry and immunology were normal.

CASE 4

This 6 year old female presented with a recent (six month) history of esotropia with diplopia. Uncorrected vision was 6/9+, N 3. She had an esotropia that measured 40° for distance and for near. Cyclorefraction was + 1.00 DS OU. Manifest refraction was plano. Coronal CT scans showed enlarged extraocular muscles. She had BMR 5.5 OU. In the early post-operative period she had a small esophoria. Biopsy results were identical to case 1.

Thyroid chemistry was minimally abnormal when performed in two different laboratories.

| | LAB 1 | LAB 2 |
|-----|--------------|---------------|
| TSH | 0.56 (0.3-5) | 0.4 (0.5-4.5) |
| T4 | 24 (12-23) | 19 (10-25) |
| T3 | 7 (3.5-6.7) | |

Antibodies – negative to Thyroglobulin (x2), Thyroid peroxidase (x2) and TSH receptor.

DISCUSSION

Strabismus associated with extraocular muscle enlargement unassociated with thyroid eye disease has been anecdotally reported by others e.g. three cases at a Jampolsky Fellows Meeting, one case by Gupta to the Paediatric Ophthalmology List Server and one case is reported in the textbook by Souza-Dias. An 8 month old girl almost identical to our Case 3 was reported by Dickson (Ophthalmology 1994;101: 1902-1907).

Our four patients had no ocular or systemic findings on history or examination to suggest thyroid eye disease. We believe that their youth and normal or near-normal blood results and lack of other features on history and examination eliminate thyroid eye disease as a credible diagnosis in our patients.

CONCLUSION

Enlarged extraocular muscles can be seen in young patients with unusual strabismus. CT scanning, muscle biopsies and systemic evaluation of similar patients may help determine the frequency of this clinical syndrome and its possible association with thyroid eye disease.