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Idiopathic orbital pseudotumours in adults*

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SUMMARY

We treated 24 cases of orbital pseudotumour from January 1981 through January 1993. The clinical characteristics and treatment outcomes were analyzed retrospectively by reviewing the medical records, radiological studies, and histological examination. All patients presented with symptoms related to the eye (proptosis, lid swelling, limited ocular motion and/or pain, chemosis, and visual disturbance), while only five patients had symptoms pertaining to the ear, nose, and throat besides the ophthalmological symptoms. Plain X-ray findings were not contributory to the diagnosis. Computed tomograms (CT) showed non-specific findings such as hypertrophy of the extra-ocular muscles and well-defined or poorly defined mass. Pathological findings were non-specific, only to reveal benign lymphoid hyperplasia and inflammatory cell infiltration with necrotizing vasculitis. Twenty-four patients were treated with high-dose steroid therapy which resulted in a significant improvement in 10 patients (42%).

Key words: orbital pseudotumour, radiological finding, histological finding, steroid therapy

INTRODUCTION

Orbital pseudotumour is an idiopathic, non-specific, space-occupying inflammation involving the intra-orbital structures (Blodi et al., 1967). Since the first description by Birch-Hirschfeld (Blodi et al., 1967; Chavis et al., 1978), its clinical implications have increased in the fields of ophthalmology and otolaryngology. Orbital pseudotumour usually presents with peri-ocular pain, exophthalmos, and proptosis. Orbital CT scans with contrast enhancement may reveal variable findings depending on the structures affected by this disease entity.

It is suggested that this disease entity may be an autoimmune disorder, like peri-arteritis nodosa and systemic lupus erythematosus (Blodi et al., 1967). Noguchi (1991) observed eosinophilic degranulation in orbital pseudotumour by immunofluorescence. However, its precise aetiology and pathogenesis remains to be elucidated. High-dose steroids or low-dose irradiation has been used for the treatment of orbital pseudotumour. Our experience with orbital pseudotumor has been analyzed in terms of clinical features and treatment outcome.

MATERIAL AND METHODS

Twenty-four patients with orbital pseudotumour treated from January 1981 through January 1993 were reviewed with respect to clinical features, radiological and histological findings, and treatment outcome. Seventeen out of 24 patients (71%) were male and seven (29%) were female. The patients' ages at presentation ranged from 23 to 78 years, with a slight preponderance in the fourth decade (8 cases: 32%).

RESULTS

Clinical features

The most common manifestation was proptosis, which was present in 16 of 24 patients (67%). Other manifestations included lid swelling, limited ocular motion, ocular pain, chemosis or visual disturbance (Table 1). As to otolaryngological symptoms and signs, facial pain and cheek swelling were present in two patients, respectively, and rhinorrhoea in one patient (Table 2). A gradual onset of symptoms (19 of 24 patients: 79%) was more common than an abrupt onset (5 out of 24 patients: 21%), if the duration of symptoms prior to the presentation was divided at three weeks. There was bilateral presentation in two patients. In 22 unilateral cases, the left orbit (13 of 22 patients: 59%) was more frequently affected than the right (9 of 22 patients: 41%). There was a slight male predominance with a ratio of 2:1. Some biological data are presented in Tables 3–4. There was no specific abnormality except for C-reactive protein and erythrocyte sedimentation rate.

Radiological findings

Plain X-rays showed no specific findings in the orbit. However, there were some associated findings in the paranasal sinuses: mucoperiosteal thickening in two patients (8%), and air-fluid level and polypoid change in one patient (4%), respectively (Table 2).

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Table 1. Ophthalmological manifestations (n=24).

symptoms or signs	n*	%
proptosis	16	67
ocular pain	14	58
lid swelling	13	54
limited ocular motion	13	54
visual disturbance	7	29
chemosis	6	25

*: The numbers are not mutually exclusive.

Table 2. Patients' data (n=24).

clinical data	n	%
ophthalmological manifestations:		
corneal reflex	23	96
pupillary reflex	23	96
otolaryngological manifestations:		
facial pain	2	8
cheek swelling	2	8
rhinorrhoea	1	4
plain X-ray findings of the paranasal sinus:		
mucoperiosteal thickening	2	8
air-fluid level	1	4
polypoid change	1	4

Table 3. Patients' quantitative biological data (n=24).

	patients	normal range
white blood cell count	9.4±4.0	4.0-10.0×10^3/mm^3
haemoglobin	13.7±1.5	12-17 g/dl
haematocrit	40.9±4.9	36-52%
platelet count	292.3±112.1	130-400×10^3/mm^3
erythrocyte sedimentation rate	22.3±19.0	up to 20 mm/h
aspartate aminotransferase (AST)	25.8±10.4	up to 40 IU/l
alanine aminotransferase (ALT)	33.6±11.2	up to 40 IU/l
thyroid function test:		
triiodothyronine (T3)	118.7±30.8	87-184 ng/dl
thyroxine (T4)	7.7±1.9	5.6-13.1 mg/dl
thyroid-stimulating hormone (TSH)	3.8±6.6	0.4-4.1 mIU/ml
T3 resin uptake	30.2±3.3	23-36%

Table 4. Patients' qualitative biological data (n=24).

	positive	negative
anti-nuclear antibody	0	24
VDRL	0	24
rheumatoid factor	0	24
C-reactive protein	24	0
anti-streptolysin-O	0	24

On CT scans with contrast enhancement of the orbit, extra-ocular muscle hypertrophy was the most common finding, which was observed in 15 of 24 patients (63%; Figure 1). Other findings included lachrymal gland swelling and well-defined or poorly-defined mass (Table 5).

Table 5. Orbit CT findings (n=24).

findings	n*	%
extra-ocular muscle hypertrophy	15	63
well-defined mass	8	33
poorly-defined mass	8	33
lachrymal gland swelling	3	13

*: The numbers are not mutually exclusive.

Treatment outcome and histological findings

Steroid therapy with prednisolone was given to 24 patients: an initial dose of 80 mg for 7-14 days and subsequent decreasing doses. A significant improvement in the symptoms and signs was observed in 10 out of 24 patients (42%), whereas 14 patients (58%) did not respond to steroid therapy. One patient who had

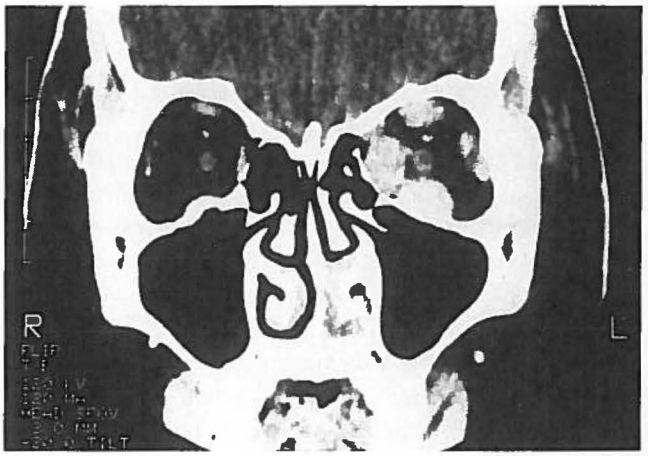

Figure 1. Coronal CT scan from a 40-year-old female patient, demonstrating hypertrophy of the extra-ocular muscles on the left side.

received steroid therapy underwent orbital decompression during steroid therapy due to progressive visual loss.

Table 6. Histological findings in non-responders to steroid therapy (n=14).

findings	n	%
perivascular infiltration of PMN leukocytes and plasma cells	7	50
lymphocytic infiltration with giant cell granuloma	5	36
perivascular infiltration of PMN leucocytes and plasma cells with myositis	2	14

PMN: polymorphonuclear

Biopsy specimens have been taken from 14 patients who showed no symptomatic improvement following steroid therapy. Histological examinations are presented in Table 6.

DISCUSSION

Orbital pseudotumour syndrome is known to be an inflammation of the intra-orbital soft tissue that represents 25% to 50% of all orbital tumours (Weisberger et al., 1985), with 16% of unilateral presentation (Fortson et al., 1980). Besides the orbit, it may involve the paranasal sinuses, larynx, clavicle, pharyngeal space, cervical spine, or may extend into the pterygomaxillary fissure (Eshaghian et al., 1981; Brazier et al., 1983; Schonder et al., 1985; Keen et al., 1986; Weisman et al., 1988). In this series, five out of 24 patients (21%) presented with otolaryngological manifestations such as rhinorrhoea, cheek swelling or facial pain. It may occur predominantly in the fourth, fifth, and sixth decades (Blodi et al., 1967; Mottow et al., 1978). Orbital pseudotumour most commonly occurred in the fourth decade, followed by the fifth and third decades. Orbital pseudotumour occurred twice as frequently in males as in females. The presenting symptoms frequently consist of proptosis, peri-ocular pain, chemosis, lid oedema, lid erythema, loss of ocular motility, and loss of visual acuity. Orbital pseudotumour may have abrupt onset in children (Mottow et al., 1978), and may accompany erosion of the cranial bone with multiple cranial nerve paralysis (Weisberger et al., 1985; Keen et al., 1986). In this series, gradual onsets were more common than abrupt ones.

For the diagnosis of orbital pseudotumour, thorough studies should be performed of the adjacent structures in the head-and-neck area. Although plain radiographs may reveal increased soft-tissue densities or radiolucent defects, they are not always pathognomonic. Orbit CT and ultrasonography may be important diagnostical tools to differentiate this disease entity from other space-occupying lesions. Orbit ultrasonography may reveal a "mottled appearance" of the orbital content and hypertrophy of the extra-ocular muscles (Chavis et al., 1978; Harr et al., 1982). Orbital CT-scans frequently show hypertrophy and contrast-enhancement of the sclera and choroid (Rowe et al., 1980; Harr et al., 1982), soft tissue density in the posterior orbit (Rowe et al., 1980; Weisberger et al., 1985) or hypertrophy of the extraocular muscles (Rowe et al., 1980; Harr et al., 1982).

In this series, hypertrophy of the extraocular muscles was more constant (15 out of 24 patients; 63%) than other radiological findings. Histological examinations may be required in selected patients with cranial nerve involvements or extensive infiltration into the surrounding soft tissue.

There are several clinical conditions that must be considered in the differential diagnosis of orbital pseudotumour (Table 7). These conditions include neoplasms of adjacent structures (such as the paranasal sinuses or the nasopharyngeal cavity), bacterial or mycotic infections of the paranasal sinuses, dermoid cyst, tuberculosis, syphilis, peri-arteritis nodosa, sarcoidosis, Wegener's granulomatosis, and thyroid ophthalmopathy.

In this series, histological examination was performed in 14 patients who were refractory to steroid therapy. The most common histological finding was perivascular infiltration of polymorphonuclear leukocytes and plasma cells in nine out of 14 patients (64%), followed by lymphocytic infiltration with giant cell granuloma in five out of 14 patients (36%).

Orbital pseudotumour may be treated with high-dose systemic steroid, low-dose radiotherapy, or orbital decompression. High-dose systemic steroid therapy is frequently effective and low-dose radiotherapy of 1,000-2,000 Gy may be effective in patients with no response to systemic administration of steroids or recurrence after systemic steroid therapy (Kim et al., 1978; Sergott et al., 1981). Special care must be paid before surgical intervention because of aggravation of the disease following surgical intervention (Blodi et al., 1967; Mottow et al., 1978). Orbital decompression and/or sinus surgery may be necessary in cases with impending blindness. In this series, 10 out of 24 patients (42%) have shown improvement by systemic steroid therapy. One patient who underwent orbital decompression due to progressive visual loss has shown no improvement following surgery. The results of this study suggest that high-dose steroid therapy may be effective in the treatment of orbital pseudotumour. In conclusion, CT scans are required for the diagnosis of orbital pseudotumour. Tissue diagnosis is indispensable when the lesion is suspicious of malignancy or when there is a poor response to steroid therapy. Orbital pseudotumour can be primarily managed with high-dose steroid therapy.

Table 7. Differential diagnosis of orbital pseudotumour.

differential diagnosis	laboratory studies and imaging
endocrine ophthalmopathy	thyroid function test
lymphoma	CBC, ESR, abdominal CT, bone marrow biopsy
orbital tumour	carotid angiography, orbital or PNS CT
Wegener's granulomatosis	BUN/creatinine, urinalysis, complement, IgA, chest PA, PNS view, renal biopsy
periarteritis nodosa	complement, BUN/creatinine, urinalysis, renal biopsy
sarcoidosis	chest PA, Kvein test, immunoglobulin, serum calcium
tuberculosis	chest PA, acid-fast bacillus culture/smear
syphilis	VDRL, TPHA, FTA-ABS
mycotic infection	fungus culture, tissue biopsy and culture
ethmoidal sinusitis	bacteriological study, PNS X-ray

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