Vol. 16, No. 4
ORIGINAL PAPER

OPHTHALMOLOGY

MEDICAL CHANNEL
OCTOBER- DECEMBER 2010

PRESENTATION OF IDIOPATHIC, NON-SPECFIC, ORBITAL INFLMMATION (PSEUDOTUMOR) - STUDY OF 46 CASES.

1- **PARTAB RAI**
FCPS
2- **SYED IMTIAZ ALI SHAH**
FCPS
3- **SYED AFTAB HUSSAIN SHAH**
FCPS
4. **AZIZULLAH JALBANI**
FCPS
5. **IMDAD ALI ANSAREE**
FCPS

1- ***Associate Professor***
Department of Ophthalmology
CHANDKA MEDICAL COLLEGE LARKANA.
2- **Head of Department of Ophthalmology**
CHANDKA MEDICAL COLLEGE LARKANA.
3- ***Associate Professor***
Head of Department of Medicine
CHANDKA MEDICAL COLLEGE LARKANA.
4. ***Assistant Professor***
Department Medicine
CHANDKA MEDICAL COLLEGE LARKANA.
5. ***Assistant Professor,***
Department Medicine
CHANDKA MEDICAL COLLEGE LARKANA.

Address for Correspondence:
DR: PARTAB RAI
FCPS
Associate Professor
Department of Ophthalmology
CHANDKA MEDICAL COLLEGE LARKANA, LARKANA
Tel: 0333-7567347
E mail- dr_partab_rai @ yahoo.com

ABSTRACT

PURPOSE OF STUDY: *To discuss clinical features, treatment and outcome of the disease.*
DESIGN: *Hospital based prospective study.*
DURATION: *From September 2001 to date.*
PLACE OF STUDY: *Department of Ophthalmology, Chandka Medical College Hospital Larkana.*
MATERIAL AND METHODS: *Our study included 46 patients with pseudotumor. From eye out patient department all patients were selected, where a specific proforma containing informed consent, patient's bio data, history, clinical examination, investigations, treatment, and follow up was filled .The disease is diagnosed by exclusion criteria, based on history, clinical features and investigations .For exclusion of systemic cause of the disease, all patients were referred to general physician. The follow-up period was carried out for at least 5 years. Only cases of nonspecific inflammatory orbital pseudotumor were included in this study.*
RESULTS: *Of these 46 cases with age range of 12 years to 58 years, 28 (60.86%) were males and 18 (39.13%) females .The disease was bilateral in11 (23.91%) cases and unilateral in 35(76.08%) cases, of which left orbit was involved in 19(41.30%) cases and right in 16(34.78%) cases. We have seen 18(39.13%) cases of myositis, 13(28.26%) cases of dacryoadenitis, 8(17.39%) cases of anterior pseudotumor, 5(10.86%) cases of diffuse pseudotumor, and 2(4.34%) cases of apical pseudotumor. The out come of our patients is that, 30 (65.21%) patients who presented earlier were successfully treated by corticosteroids and 3(6.52%) recurrent, steroid resistant, biopsy proven cases were treated by low-dose orbital radiotherapy. Due to late presentation of 13(28.26%) patients complications were seen, exposure keratopathy 4(8.69%) cases, rectus muscle paresis 3(6.52%) cases, secondary glaucoma 2(4.34%) cases, optic atrophy 2(4.34%) cases and frozen orbit 2(4.34%) cases. The last 6 (13.04%) patients developed complete visual loss in the end.*
CONCLUSION: *Early presentation of patient, during initial stage of the disease will result in decreased risk of disease extension and ocular complications.*

KEY WORDS: Idiopathic orbital pseudotumor —— 46 cases study

INTRODUCTION

Orbital pseudotumor is defined as non-specific, idiopathic inflammatory process in the orbit or eye. Unlike cancerous tumors, it cannot invade other tissues or spread elsewhere hence it is called pseudotumor. Among orbital disorders, pseudotumor, after Grave's disease and lymphoproliferative disease, is a common ophthalmologic disease, about 4.7 to 6.3% [1]. The cause is unknown and usually unilateral (90%). The disease usually occurs in adults but may also affect children. Pediatric orbital pseudotumor encompasses about 6% to 16%. In children, there is a higher incidence of bilateral orbital involvement without evidence of underlying systemic disease. The disease may present acutely, sub-acutely, or chronically in one orbit or may occur bilaterally and may be recurrennt[2]. Nonetheless, immune response cells, in particular lymphocytes and different stages of local fibrosis and neovascularisation are present in histopathological specimens of pseudotumor in newly formed connective tissue. Indeed, the relative proportion of inflammatory cells with respect to collagen fibers is used to differentiate the pseudotumor

TABLE 1

SHOWING ANATOMIC LOCALISATION, NUMBER OF PATIENTS, SEX, AGE RANGE AND LATERALITY OF PATIENTS WITH PSEUDOTUMOR. (N =46).

S No	Cause	No of Patients	Male	Female	Age range	Laterality
01	Myositis	18	11	7	12y-46y	Bil:5 Rt:6 Lt: 7
02	Dacryoadenitis	13	6	7	14y-58y	Bil:3 Rt:4 Lt:6
03	Anterior pseudotumor	8	5	3	22y-53y	Bil:2 Rt:4 Lt:2
04	Diffuse Pseudotumor	5	3	2	18y-57y	Bil:1 Rt:1 Lt: 3
05	Apical Pseudotumor	2	2	0	27y-44y	Rt:1 Lt:1
	Total	**46 cases cases**	**27(58.69%) cases**	**19(41.30%) 8years.**	**12-5**	**Bil: 11(23.91%) Rt: 16(34.78%) Lt: 19(41.30%) Cases**

KEY: y= years, RT= right, Lt=left, Bil= bilateral

into the more frequently occurring inflammatory type versus the rare fibromatous type. Perivascular Inflammatory changes and secondary inclusion of vessels occurs rather more frequently, which testifies to the somewhat "atypical" histological pattern of the pseudotumor[3, 4]. .The disease is classified according to the site of inflammatory process which may be localised or diffused. Localised forms of inflammation involve the anterior or posterior (apex) orbit, extraocular muscles and lacrimal gland. The severe proptosis can lead to exposure keratopathy, diplopia, secondary glaucoma and blindness. The radiologic evaluation consists of CT scan and MRI of orbit. The imaging findings, correlated with the clinical findings, allow a diagnosis in most cases and obviate the need for a biopsy. For elucidation and confirmation of the suspected clinical diagnosis, the CT scan is the preferred method because of the inherent contrast by different attenuation values of the orbital fat, muscle, bony structures, and air in the adjacent paranasal sinuses. Extraorbital extension, however, especially to the cavernous sinuses is better delineated on MRI imaging. At times orbital fatty infiltration and perineuritis are better delineated on fat suppression T1- weighted MR images than on CT scans. [5,6]

Treatment – 60 to 80mg of oral prednisone per day. In improving patients, their steroids are tapered off after 2-3 weeks at a rate of 5 – 10 mg per week over 2 to 3 months period. Failure to respond requires orbital biopsy. If biopsy shows benign process, then low-dose orbital radiotherapy (between 1000-3000 rads) is advised and if biopsy shows lymphoproliferative disorder, then appropriate treatment is instituted. Cyctotxic agents (Cyclophosphamide, low dose cyclosporine) have been used in case refractory to corticosteroids and radiotherapy**.** [7].

MATERIAL AND METHODS

Prospectively we studied 46 patients of pseudotumor aged between 18 years to 58 years from September 2001 to date at department of ophthalmology, Chandka medical college hospital larkana .In each case , patients name ,age ,sex ,occupation ,address , detailed history, examination, and was noted on a specific proforma . The diagnosis of this idiopathic disease was based on the exclusion of other causes of orbital lesions by history, clinical features of eye and underlying systemic disease, and relevant investigations where necessary like complete blood cell count with morphology, erythrocyte sedimentation rate (ESR), total and differential leukocyte count (TLC,DLC), fasting blood sugar (FBS), serum- thyroid stimulating hormone (TSH), thyroxin(T3), thyronine(T4), rheumatoid arthritis (RA) factor, serum angiotensin converting enzymes (ACE), serum calcium level (S-Ca), anti-nuclear antibodies (ANA), anti-neutrophilic -cytoplasmic antibodies (ANCA),anti-smooth muscle antibodies (ASMA),C-reactive proteins (CRP), , enzyme linked immunosorbent assay (ELISA) ,Mantoux test (MT), polymerase chain reaction (PCR), Gram's staining and culture of purulent discharge of lids ,conjunctiva, blood culture and sensitivity, radio / imaging techniques like ultrasound abdomen, x-ray orbits / chest / sinus, computerised tomography scanning (CT Scan) of orbits, magnetic resonance imaging (MRI) of orbits, ocular B-scan, and orbital soft tissue biopsy in recurrent cases resistant to steroid therapy .For exclusion of systemic cause of the disease, all patients were referred to general physician . All the patients were treated by systemic corticosteroids, analgesics, topical eye antibiotic drops and ointment to prevent secondary ocular infection and exposure keratopathy. The steroids resistant and orbital soft tissue biopsy proven pseudotumor patients were treated by local orbital radiotherapy. The follow-up period was

TABLE 2
SHOWING SPECIFIC ANATOMICAL LOCALIZATION, PRESENTATION, CLINICAL EXAMINATION, INVESTIGATIONS AND TREATMENT OF PATIENTS WITH PSEUDOTUMOR. (N = 46)

S No:	No. of Patients	Anatomic location	Presentation	Clinical Examination= ocular and orbital)	Investigations= orbital	Treatment
1	18 (39.13%)	Myositis	Pain on eye movement, .localized eye redness	BCVA= 6/6, painful decreased EOM, localised injection and chemosis.	CT,MRI=muscle irregularly enlarged, swelling of tendon, local episcleral and scleral swelling ,fusiform enlargement of whole muscle B-scan= increased extraocular muscle size.	Topical dexamethasone 1% eye drops QID for 7days then tapered over in 6 weeks. +1+2+3+4
2	13 (28.26%)	Dacry-oadenitis.	Pain, redness and swelling in the superotemporal region of the orbit.	BCVA= 6/6, lateral swelling, S-shaped lid deformity, tenderness, pouting of lacrimal ducts, chemosis, localized injection.	CT, MRI= irregular, swelling of lacrimal gland and adjacent tissues. B-scan= local swelling with increased Tenon's space.	Topical dexamethasone 1% eye drops QID for 7days then tapered over in 6 weeks.+1+2+3+4
3	8 (17.39%)	Anterior pseudo tumor.	Moderate pain, redness, swelling of eye with mild dimness of vision	BCVA=6/9 -,6/6P , diffuse injection and swelling of lids, chemosis, and painful decreased EOM.	CT, MRI= Anterior enhancement with irregular margins close to sclera .Irregular extension along optic nerve. Decreased fat density. B-Scan= sclerotenonitis with T-sign.	1+2+3+4+5
4	5 (10.86%)	Diffuse pseudo tumor.	Moderate to severe pain, redness, swelling of eye with mild to moderate dimness of vision.	BCVA=6/12 -6/9, diffuse injection and swelling of lids, chemosis, and painful decreased EOM.	CT, MRI= Diffuse enhancement with decreased fat density. B-Scan= T-sign.	1+2+3+4+5
5	2 (4.34%)	Apical pseudo tumor.	Severe pain, redness, swelling of eye with moderate to severe dimness of vision	BCVA=6/60-6/12, decreased EOM, chemosis, mild to moderate proptosis.	CT,MRI= Apical irregular infiltration extending along with muscle and optic nerve	1+2+3+4+5

1. Oral flurbiprofen 100 mg BD for a week.
2. Oral prednisone1 mg/kg/ day for a week then tapered over 5-10 mg /week over 8-12weeks period.
3. Oral omeprazole 40 mg /day till steroids stopped
4. Topical antibiotic ciprofloxacin eye drops and eye ointment for 1-2 weeks.
5. 3 ml intravenous dexamethasone (4mg/ml) / day for 5days then followed by oral prednisone1 mg/kg/ day for a week then tapered over 5-10 mg /week over 8-12weeks period

BCVA=Best corrected visual acuity.

EOM=Extraocular movements

PHOTOGRAPH .1



Photograph of 22 years old female showing right eye lateral rectus myositis.

PHOTOGRAPH.2



Photograph of 38 years old female showing left eye dacryoadenitis.

PHOTOGRAPH .3



Photograph of 34 years old female showing left eye anterior pseudotumor.

PHOTOGRAPH. 4



Photograph of 28 years old female showing left eye diffuse pseudotumor.



The photograph of same patient after treatment

PHOTOGRAPH .5



Photograph of 18 years old female showing bilateral apical pseudotumor (Before treatment).



The photograph of same patient after treatment showing left eye medial rectus paresis and divergent squint.

PHOTOGRAPH .6



Photograph of 32 years old male showing left eye steroids resistant diffuse pseudotumor with local radiotherapy landmarks.

carried out for at least 5 years. Only cases of nonspecific inflammatory orbital pseudotumor were included in this study.

RESULTS

Of these 46 cases with age range of 12 years to 58 years, 28 (60.86%) were males and 18 (39.13%) females .The disease was bilateral in11 (23.91%) cases and unilateral in 35(76.08%) cases, of which left orbit was involved in 19(41.30%) cases and right in 16(34.78%) cases. According to specific anatomical localization of pseudotumor we have seen 18(39.13%) cases of myositis, 13(28.26%) cases of dacryoadenitis, 8(17.39%) cases of anterior pseudotumor, 5(10.86%) cases of diffuse pseudotumor, and 2(4.34%) cases of apical pseudotumor. (Please see table 1). After diagnosis, the patients with severe acute pseudotumor were treated by 3 ml intravenous dexamethasone (4mg/ml) / day for first 5days then followed by oral steroids. The patients with mild to moderate pseudotumor were treated by oral prednisolone 1mg/kg/day for first 7days, then oral steroids were tapered off at a rate of 5-10 mg / week over 2 to 3 months period with oral omeprazole 40 mg/day (till steroids continued), oral flurbiprofen 100 mg twice a day (till eye pain relieved), topical ciprofloxacin 3% eye antibiotic drops and ointment for first 1 to 2 weeks. (Please see table 2). The out come of the our 46 patients is that, 30 (65.21%) patients who presented earlier were successfully treated by corticosteroids and 3(6.52%) recurrent steroid resistant cases who also presented earlier , after proving pseudotumor by orbital soft tissue biopsy were treated by low-dose orbital radiotherapy (between 1000-3000 rads) . Due to late presentation of 13(28.26%) patients complications were seen, exposure keratopathy 4(8.69%) cases, rectus muscle paresis 3(6.52%) cases, secondary glaucoma 2(4.34%) cases, optic atrophy 2(4.34%) cases and frozen orbit 2(4.34%) cases. The last 6 (13.04%) patients developed complete visual loss in the end.

DISCUSSION

We have seen about 845 cases of various types of orbital diseases from September 2001 to date, of which 46 (5.44%) cases of pseudotumor were picked up which account similar number of patients as seen by *Henderson John Warren.*[1] et al 5.2 %.The clinical presentation of orbital pseudotumor was chiefly determined by the degree of inflammatory response and the particular orbital tissues involved. The inflammation may be localized or diffused. Localized forms of inflammation involved the anterior or posterior (apex) orbit, extraocular muscles, and lacrimal gland. Here each form of disease is discussed separately. We have seen 18(39.13%) cases of ***myositis*** with clinical features of acute or subacute onset of painful extraocular movements, diplopia, ptosis and swelling of the lid, localized chemosis and injection over the affected muscle and limitation of extraocular movements. CT scan showed relatively diffuse enlargement of the involved extraocular muscle with slightly irregular margins. The most frequently affected muscles were the superior complex, medial and lateral rectus. (Please see photograph 1) The major differential diagnosis of this condition is Grave's orbitopahty. However dysthyroid myopathy is usually painless in onset, asymmetric, slowly progressive, and associated with systemic features [8.] We have seen 13(28.26%) cases of dacryoadenitis with age range from 16 to 52 years, and more in females 7(15.21%) cases than males 6(13.04%) cases. The typical presentation of the acute disorder consisted of pain, tenderness, and injection of the temporal part of upper lid and of conjunctival fornix with an associated palpable lacrimal gland, S- shaped deformity of the upper lid, and pouting of the lacrimal ducts noted on slit lamp examination of the fornix. (Please see photograph 2) This group showed minimal proptosis, and downward and inward displacement of the globe. Lacrimal gland inflammation may also be subacute or chronic, evolving over weeks to months and presenting with a painless lacrimal fossa mass. The CT scan showed enhancement and enlargement of the lacrimal gland. The differential diagnosis of this idiopathic nonspecific dacryoadenitis includes viral and bacterial dacryoadenitis, specific dacryoadenitis such a sarcoidosis and Sjogren's disease, neoplasia, reactive lymphoproliferative disorders and granulomatous dacryoadenitis. [9] We have seen 8(17.39%) cases of anterior pseudotumor, which primarily involved tenon's capsule, episclera, and sclera. The main symptoms and signs in these patients were pain, redness and swelling of the eye and ocular adnexa, proptosis, lid swelling, and injection. (Please see photograph 3) .The age of patient ranged from second to fifth decade. The CT scan showed anterior orbital infiltration intimately related to the globe, producing scleral and choroidal thickening with obscuring of the junction of the globe and optic nerve and variable extension along its sheath. On B-scan ultrasonography, there was an irregular uniform density with anterior orbital infiltrate, sclerotenonitis with accentuation of tenon's capsule and doubling of the optic nerve shadow (T-sign).The differential diagnosis of anterior orbital inflammation includes, orbital cellulitis, local ocular inflammation (scleritis, uveitis), systemic lesions like rhabdomyosarcoma, leukaemia, lymphoma and metastatic neuroblastoma. In our study 5(10.86%) cases of diffuse pseudotumor presented by similar signs and symptoms of anterior orbital inflammation in addition with greater severity of limitations of ocular movements (Please see photograph 4). CT scan showed soft tissue infiltration involving the entire orbit, extending from the apex to the posterior margin of the globe. The optic nerve and extraocular muscles are obscured to a variable extent. B-scan ultrasonography showed doubling of the optic nerve shadow (T-sign), sclerotenonitis and orbital infiltration.[10]. 2(4.34%) patients were seen with posterior apical pseudotumor. They presented with a typical orbital apical syndrome of pain, proptosis restricted extraocular movements and decrease vision (Please see photograph 5). The CT scan showed an irregular infiltration of the apex of the orbit with extension along the posterior portion of extraocular muscle or the optic nerve. The differential diagnosis of this condition includes optic neuritis, apical tumefaction, and Tolosa - Hunt syndrome[11]. Like study of Mottow LS [12] and Mottow-Lippa L [13] et al, paediatric pseudotumor was seen during the first and second decades of life. Also there was more bilateral orbital involvement with mild anterior uveitis and optic disc edema in the paediatric population 7(15.21%) cases than adults and older 4(8.69%) cases. Weber AL et al [14] in their study and in our study we have noticed that trauma in 4(8.69%) cases and upper respiratory tract infection in 3(6.52%) cases preceded the onset of pseudotumor and there was eosinophilia in the peripheral blood in 11(23.91%) cases. In our study 30 (65.21%) patients who presented earlier were successfully treated by corticosteroids. Severe 3(6.52%) recurrent , steroid resistant cases who also presented earlier , which after proving pseudotumor by orbital soft tissue biopsy were treated by low-dose orbital radiotherapy (between 1000-3000 rads). (Please see photograph 6). Due to late presentation of 13(28.26%) patients the complications were seen, exposure keratopathy 4(8.69%) cases, rectus muscle paresis (Please see photograph 5) 3(6.52%) cases, secondary glaucoma 2(4.34%) cases, optic atrophy 2(4.34%) cases and frozen orbit 2(4.34%) cases. The last 6 (13.04%) patients developed complete visual loss.

CONCLUSION

Early presentation of patient during initial

stage of the disease with immediate diagnosis and treatment will reduce the risk of disease extension and ocular complications.

REFERENCES:

1. Warren HJ. Inflammatory orbital tumors, in orbital tumors by Henderson 3rd ed, 1994, Raven press, New York, pp 1923-1939.
2. Snebold Neal G. Non-infectious orbital inflammations and vasculitis In principles and practice of ophthalmology by Albert and Jakobeic vol 3,1994,WB Saunders company, London,pp.1923-1939.
3. Mombaerts I, Goldschmeding R, Schlingemann R O, Koorneef L .What is orbital pseudotumor? Surv Ophthalmol, 1996; 41 (1):66-78.
4. Shi J T; An YZ,Sun XL, Li B. Clinical and pathologic analysis of orbital pseudotumor.Zhonghua Yan Ke Za Zhi , 2003;39(2):81-6.
5. Kanski Jack J. The orbital cellulitis.In clinical ophthalmology by Kanski 6th edition 2008, Butterworth –Heinemann company, London, pp558-588).
6. Rootman J, Robertson William, Lapointe Jocelyn S .Inflammatory diseases. In the diseases of the orbit by J Rootman, 2nd ed 2003, JB Lippincott company, London, pp.455-499.)
7. Garrity JA, Kenner dell JS, Johnson BL. Cyclophosphamide in the treatment of orbital vasculitis .Am J ophthalmol 1986; 102: 97-103
8. Rubin PA, Foster CS. Etiology and management of the idiopathic orbital pseudotumor inflammation. Am J Ophthalmol 2004; 138:1041-1043.
9. Rootman J, White VA, Connors JM, Gascoyne RD. Lymphoproliferative, leukaemia, and histiocytic lesions of the orbit. In the diseases of the orbit, by J Rootman, 2nd ed 2003, JB Lippincott company, London, pp385-416.
10. Rootman J, Nugent RA. The classification and management of acute orbital pseudotumors. Ophthalmology 1982; 89:1040-8.
11. Wasmeier C, Pfadenhauer K, Rosler A. Idiopathic inflammatory pseudotumor of the orbit and Tolosa Hunt Syndrome-are the same disease ?J Neurol 2002;249:1237-1241.
12. Mottow LS, Jakobeic F A. Idiopathic inflammatory orbital pseudotumor in childhood .l. Clinical characteristics. Arch Ophthalmol 1978; 96:1410-1417.
13. Mottow-Lippa, Jakobeic F A, Smith M. Idiopathic inflammatory orbital pseudotumor in childhood .ll. Results of diagnostic tests and biopsies .Ophthalmology 1981; 88:565-574.
14. Weber AL, Romo LV, Sabates NR.Pseudotumor of the orbit clinical pathologic and radiologic evaluation. Radiol Clin North Am 1999; 37:151-168.