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DIFFERENT ASPECTS OF OROFACIAL PAIN (PART III)
ORBITAL AND PERIORBITAL PAIN

RAZLIČITI ASPEKTI OROFACIJALNOG BOLA (TREĆI DEO)
ORBITALNI I PERIORBITALNI BOL

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Abstract

The sensation of ophthalmic/orbital pain may result from both direct or indirect stimulation of pain fibers and their anatomically related structures. Noxious stimuli may affect the trigeminal nerve and its branches, also the facial, glossopharyngeal, and vagal nerves. Localization of pain may be difficult because of the convergence of pain information from smaller branches to major trunk (i.e. ocular pain experienced as headache in acute angle closure glaucoma in contrast to eye pain, which is referred from and confused with an upper molar abscess). The sensation of pain may be experienced as sharp, carried by small myelinated A delta fibers, and dull, aching pain, carried by small unmyelinated C fibers. In general, the origin of the pain is classified as deriving from the globe, orbit, head, or, when no source can be defined, as functional.

Key words
orbital, periorbital and ophthalmic pain, diagnostics, treatment

Ključne reči
orbitalni, periorbitalni i oftalmički bol, dijagnostika, lečenje

INTRODUCTION TO NEUROANATOMY AND PATHOPHYSIOLOGY

Sensory Innervation of the Eye and Orbit

The globe itself is pierced by the long and short posterior ciliary nerves that branch directly from the nasociliary nerve, a branch of the opthalmic division of the trigeminal nerve (V1). The cornea and sclera are richly endowed with pain fibers in contrast to the conjunctiva, which is only sparsely innervated. The uveal tissue (iris, ciliary body, and choroid) is also substantially innervated.

The orbit is innervated by the opthalmic (V1) and maxillary (V2) divisions of the trigeminal nerve. The frontal branch of V1 divides into three branches: supraorbital (supplies the forehead, scalp, upper lid, and frontal sinus), supratrochlear (supplies the medial upper lid and conjunctiva), and frontal that supplies lateral upper lid and lateral upper orbit [1, 2, 3]. The lacrimal branch of V1 supplies the lacrimal gland and contiguous skin and conjunctiva; it anastamoses with the zygomatic branch of V2 and carries postganglionic parasympathetic inervation to the lacrimal gland, by way of the sphenopalatine ganglion and sympathetic fibers. In addition to the globe, the nasociliary nerve supplies the sphenoid and ethmoid sinuses, nasal mucosa, and medial and inferior orbit. It branches into the infratrochlear nerve, which supplies the inferior lid and infratrochlear area. The zygomatic branch of V2 supplies the temporal skin and lateral inferior lid and cheek by way of the temporal and facial branches; the infraorbital branch of V2 supplies the lower eyelid, mid face, and upper lip [1, 2].

The cavernous sinus transmits to V1 and V2 branches to supply intracranial structures, which can be the sites of referred ocular and adnexal pain; meningeal branches of V1 supply the tentorium cerebelli, falx cerebri, cribriform plate, and both wings of the sphenoid. The internal branches of the zygomatic nerve supply the dura mater and the vessels of the middle cranial fossa.

Mechanism of Ocular, Orbital and Periorbital Pain

When noxious stimuli involve mucosal structures, vasoactive amines and kinines (i.e. substance P) could be formed, which causes pain to be perceived both locally and centrally. Substance P, a neurotransmitter that participates in the pain response, is transmitted both localy and distaly, and converts information that the sensory cortex cannot distinguish if it originate from local or from more remote (uninvolved) structures [3], inflammation (i.e. sinusitis) or an

expanding mass (i.e. adenocarcinoma of the lacrimal gland) that may produce pain looking like it originates from the globe or orbit Cervical spondylitis may cause a referred orbital or retro-orbital pain [2] because the cervical branches of the trigeminal nucleus in the spinal tract converge to V1.

DIAGNOSIS AND MANAGEMENT OF SYNDROMES BY ANATOMIC LOCATION

Ocular

Keratitis: Mechanical or inflammatory disturbance of the cornea which is abundantly innervated, results in a severe noxious response. The pain is perceived only if the epithelium is denuded or the stromal nerves are locally disturbed, because the nerves run through the corneal stroma. Trauma, infection, chemical irritation, exposure, and dry eye might be the underlying cause. Symptoms of a foreign body sensation may be early indicators of keratitis, together with photophobia and lacrimation; whereas intense pain is the hallmark of more advanced keratitis (i.e. bacterial corneal ulcer), intensive topical antimicrobial agents are the treatments of choice for bacterial keratitis. Noninfectious irritation may require irrigation, foreign body removal, lubricants, or steroids.

Scleritis: Inflammatory processes may involve anterior or posterior sclera and may be necrotizing or no necrotizing. The underlying cause may be collagen-vascular disease or idiopathic. Anterior scleritis usually presents with intense ocular pain, whereas posterior scleritis may present as orbital pain with or without pars planitis (uveitis intermedia) or exudative retinal detachment. Treatment may include judicious use of topical and sometimes systemic steroids; occasionally antimetabolites and mucolytics are used.

Glaucoma: The classic triad of a red eye, mid-dilated pupil, and corneal oedema with elevated intraocular pressure is the hallmark of acute angle closure glaucoma. Symptoms of severe ocular pain associated with severe headache are typical; pain may radiate around the orbit, including sinuses and teeth, and be severe enough to result in mistaken dental extraction [4] or a course of antibiotics. The noxious response also may be so severe to cause nausea and vomiting. Breaking the attack with topical drops and definitively either with laser or surgical iridotomy or iridectomy is the mainstay of treatment.

Ischemic Ocular Syndrome: Carotid insufficiency may lead to anterior or posterior segment ischemia that results in corneal oedema, chemosis, rubeosis, retinal hemorrhages, and neovascularization. More localized ischemia may be caused by iatrogenic manipulation, (i.e. strabismus surgery on more than two muscles and scleral buckling surgery for too tight and encircling band). Topical steroids, atropine, cryosurgery, laser photocoagulation, carotid endarterectomy or reanastamosis [7] and corrective ocular surgery may be options.

Aesthenopia: Painful vision may result from excessive tone in the ciliary body, typically from an uncorrected refractive error. The symptoms usually are exacerbated when focusing for long periods at reading distances. Headache or brow ache, the chief complaint, could be relieved with proper corrective lenses.

Orbital

Orbital Cellulitis: Infection of the orbital tissues may result from contiguous spread of organisms that originate from the lid, sinuses or direct infiltration of a foreign body. When the infection begins in the tissues of the lid, it is called preseptal cellulitis and can present with a painful, sore lid but no eye pain; treatment is usually with oral antibiotics. The infection that crosses deeper of plane of the orbital septum produces orbital cellulitis. This condition can present as a dull ocular and facial pain early on and become quite severe, especially if subperiosteal or orbital abscess develops [5]. The pain may be exacerbated by palpation and eye movement (or attempted movement). At the more advanced stages vision may be compromised as well due to compression on the optic nerve or its vascular supply Treatment is with intravenous antibiotics and surgical drainage of abscesses or infected sinuses. It is imperative that the infection be eradicated quickly because further spread of the infection can lead to meningitis or cavernous sinus thrombosis.

Pseudotumor of orbita: This is a spectrum of inflammatory syndromes of idiopathic origin. Developing acutely or chronically, the orbital tissues become edematous, engorged from inflammatory cells and their mediators. Pain, diplopia, proptosis, chemosis, and conjunctival infection are typical presenting symptoms. The inflammation may be classified as anterior, diffuse, myositic, or lacrimal. When the superior orbital fissure or cavernous sinus is involved, it is called Tolosa-Hunt Syndrome and presents as painful ophthalmoplegia. If the optic nerve is involved and vision is compromised, the term orbital apex syndrome is used. Contrast CT or MR imaging with fat suppression technique aid in the diagnosis, and rapid response to systemic steroids helps to further establish the diagnosis. If is important to rule out lymphoma, local tumors, and metastases as infiltrative cause when little or no response to steroids is observed [7].

Trochleas: A variant of orbital pseudotumor, trochleitis or superior oblique tendonitis may present as dull aching localized to the superior - nasal, orbit, with exquisite point tenderness and exacerbation by eve movement [11]. This condition may arise secondarily from contiguous sinusitis, prior surgery, trauma, and rheumatoid arthritis. Aside from treating any underlying causes, the condition responds to nonsteroidal anti-inflammatory drugs (NSAIDs) and steroids either injected locally or given systemically.

Optic neuritis: Inflammation of the optic nerve may occur as an isolated disorder or may be associated with demyelinatmg disease. Presenting symptoms are decreased vision associated with retroocular pain, especially upon movement. The initial evaluation typically entails MR imaging of the brain to rule out demyelinating plaques, followed by pulsed steroid treatment (methylprednisolone 1 g intravenously daily for 3 days), and followed by 60 mg to 80 mg daily until vision returns. The pain may precede vision loss and often resolves before vision returns.

Tumor: Infiltration of the orbit by neoplastic and no neoplastic tumors may cause orbital pain but most often

presents as progressive pro-ptosis with or without diplopia before pain is felt. LymphoprolIferative disorders, nasopharyngeal carcinoma, lacrimal gland carcinoma, rhabdomyosarcoma, neuroblastoma, vascular tumors, and secondary metastatic tumors may be the offending cell types. Chemotherapy and adjunctive radiation treatment may aid greatly in tumor regression [7].

Head

Some remote syndromes that involve vascular and neuronal structures, the sinuses, ears, and teeth are important causes of ophthalmic pain.

Vasculopathic: Temporal arteritis is a serious disease that affects elderly individuals, usually in their seventh to ninth decades, but may occur in younger individuals, The underlying cause is an autoimmune destruction within the media of the affected arteries by invasion of giant inflammatory cells, which causes obliteration of the lumen and consequent necrosis of tissues in their distribution. Classically presenting symptoms and signs are unilateral visual loss transiently or permanently associated with throbbing pain on the ipsilateral side of the head and sometimes scalp tenderness, jaw claudication, dental pain, and otalgia. The variant of polymyalgia rheumatica may involve proximal limb-girdle arthralgias. Visual loss is a consequence of central retinal artery occlusion, ophthalmic artery occlusion, or necrotizing ischemic optic neuropathy. Rarely, homonymous hemianopsia may result if the posterior cerebral artery is involved and occipital lobe infarction occurs. Diagnosis is made clinically and corroborated by laboratory tests (e.g., high Westergren erythrocyte sedimentation rate, elevated C-reactive protein and fibrinogen levels). Definitive diagnosis is made by temporal artery biopsy usually within 2 weeks, although skip areas may occur. The mainstay of treatment is high-dose corticosteroid treatment primarily to prevent disease from affecting the contralateral eye, which may occur within 6 months in more than 75% of cases.1 Slow titration of treatment, usually over 1 year, using clinical and laboratory markers is key in suppressing the inflammation. Even upon eventual withdrawal of steroid therapy, relapses may occur.

Carotid occlusive disease may cause unilateral headache and neck and facial pain, some-times localized to the orbit. Ocular signs may include rubeosis iridis and retinal hemorrhages. Differential diagnosis includes temporal arteritis, which requires careful evaluation. Carotid Doppler studies and MR angiography help to establish the diagnosis. Unless ulcerating plaques are suspected (i.e. transient ischemic attacks, amaurosis), anticoagulation usually suffices to apply prophylaxis against stroke; otherwise carotid endarterectomy may be indicated for severe disease [7, 8, 9].

Neurogenic: Herpes zoster ophthalmicus is a latent reactivation of dormant varicella zoster infection from childhood that affects the V1, branch of the trigeminal nerve. Pain typically precedes the vesicular eruptions by 2 to 3 days and may be quite severe. Symptoms of burning, aching, and throbbing may be felt along any part of the trigeminal nerve. When the nasociliary branch of is affected, vesicles on the nose, known as Hutchinson's sign, may portend severe ocular inflammation that can include keratitis, uveitis, and glaucoma. Acute intervention with antitherpetic agents (e.g. acyclovir) may shorten the course of the disease, reduce pain, and prevent scarring over the affected dermatone (skin tightening). Almost as debilitating as the acute infection, postherpetic neuralgia may appear or persist several months after the initial presentation [8]. It can affect approximately 10% of patients and increases in frequency with the age of the individual The pain, which is exquisite at the slightest touch, improves gradually but residual hypoesthesia and hyperalgesia may persist for more than 1 year. Antiviral treatment does not prevent postherpetic neuralgia but may shorten its course. The mainstay of treatment includes analgesics, antidepressants, aloe lotions, and capsaicin (Zostrix). Capsaicin diminishes neurogenic pain by depleting substance P from peripheral nerve endings; care is needed to avoid direct contact with the eyes [5].

Trigeminal neuralgia, also known as "tic douloureux", is an idiopathic disorder that presents as paroxysmal severe pain often characterized as razor sharp, cutting, piercing, or like a tooth fracturing [3]. The pain may begin along one branch and spread over the entire distribution of that branch or neighboring branch.

Greater occipital neuralgia, also known as **occipital neuritis**, is believed to result from inflammation of the greater occipital nerve at the base of the skull or entrapment of sensory roots [2]. The presentation of occipital pain radiating to the eye, temple, and face probably stems from the close association of the greater occipital nerve and the spinal tract of the trigeminal nerve. Treatment is usually with local heat, NSAIDs, and sometimes locally injected anesthesia and steroids.

Raeder's paratrigeminal syndrome is a trigeminal neuralgia coupled with oculosympathetic palsy (Horner's syndrome) and it involves multiple cranial nerves (III, IV, V, or VI)-type 1 or isolated-type 2. Type 1 requires neuroimaging to rule out a parasellar mass [1]. Type 2 has a migrainous component that rarely may be associated with fibromuscular dysplasia and carotid dissection. Pain varies in duration from hours to weeks to months and in cluster periods 6 to 8 weeks, sometimes exactly 12 to 24 hours apart. Each episode may last 10 to 120 minutes and is characterized by severe unilateral frontal or frontotemporal pain associated with rhinor- rhea, lacrimation, conjunctival hyperemia and chemosis, and ipsilateral Horner's syndrome. Treatment usually includes aborting the migrainous attack and then breaking the cycle with systemic corticosteroids/ ergot alkaloids, and lithium chloride [9].

Migrainous: Migraine headaches often present as unilateral or bilateral ocular pain. The headache classically involves a preceding visual aura, photophobia, nausea, and vomiting and is usually localized but may be generalized. The events leading up to the attack involve platelet aggregation and consequent ischemia followed by reactive vasodilation (which in fact causes the headache). Management is by abortive or preventive treatment. Typical cases (e.g., new onset in the elderly, headache preceding visual symptoms, severe associated neurologic deficits) should be evaluated with neuroimaging [8].

Sinusitis: Acute purulent sinusitis presents as a constant,

dull aching or throbbing, which may be severe at times and exacerbated by dependent head positioning. The pain is usually localized over the affected sinus but may be referred to other structures in the head depending on which nerves are simultaneously involved.[11] Maxillary sinusitis is usually localized to the cheek but may be referred to the forehead, retroorbital, or occipital regions. Because the posterior ethmoid and sphenoid sinuses are innervated by V2 branches, the referred pain is similar to that in maxillary sinusitis. Frontal sinusitis may be localized over the forehead and deeper within the head due to innervation by branches of V1, which supply the frontal sinus and anterior cranial fossa dura. Anterior ethmoidal sinusitis may be localized to the side of the nose or medial orbit and referred to the temple by way of the nasociliary anastamosis with the zygomatic nerve by way of the frontal nerve. Chronic sinusitis is usually the cause of dull aching or pressure feeling and headaches. Treatment is with broad-spectrum oral antibiotics and, in severe cases, intravenously. Progression to orbital cellulitis may occur at any time and is heralded by ltd edema, lacrimation, and orbital pain. The differential diagnosis includes cluster headaches and mass lesions, which affect the cavernous sinus and sphenopalatine ganglion [9, 12, 13].

Chronic sinusitis may lead to the formation of a mucocele, which is caused by destruction of the sinus outflow tract. The frontal or ethmoid sinuses are most often affected, and when the mucocele protrudes into the orbit, diplopia and pain may result [10]. Surgical reconstruction of the orbit and concomitant antibiotic administration may be indicated in such cases.

Otologic: The ear receives sensory innervation from cranial nerves V, VII, IX, and X and from cervical roots 2 and 3 (C2 and C3). Initially otitis media may present locally as sudden, sharp, excruciating pain or a dull ache. In severe cases, the pain may progress to the eye, suggesting petrositis, which classically presents as orbital pain, otalgia, and aural discharge. Involvement of the petrous apex and cranial nerve VI, which travels through Dorello's canal, heralds the development of Gradenigo's syndrome, which classically manifests as diplopia, otalgia with discharge, and orbital pain [7]. Nasopharyngeal carcinoma may present with otitis media, otalgia, and cavernous sinus syndrome with multiple cranial neuropathies and is sometimes confused with Gradenigo's syndrome. Treatment of petrositis is with broad-spectrum intravenous antibiotics; nasopharyngeal carcinoma re-quires a surgical or oncologic consultation.

Dental

Inflammation of the dental nerves caused by intrinsic disease of the tooth or gum may produces symptoms of lacrimation, photo-phobia, or conjunctival injection and headaches, mimicking symptoms of trigeminal neuralgia, cluster headache, and sinusitis [2]. Treatment is by dental consultation, with appropriate referral to an endodontist, oral surgeon, or periodontist.

Miscellaneous

Cellulitis, infection of the facial skin with group A hemolytic streptococcus induces a localized pain that may spread according to the severity of the infection. Local signs include skin oedema and sharply demarcated erythema and may be associated with systemic signs of fever, chills, and malaise. Treatment is with oral antibiotics, although severe cases and those in the pediatric age group may require intravenous therapy.

Osteomyelitis: Direct extension of local infections in the nose, teeth, and sinuses and hematogenous dissemination may lead to osteomyelitis. Pain is usually felt locally, although it may be referred to remote sites in severe cases. Treatment is with a long course of intravenous antibiotics [5].

Temporornandibular joint: Temporomandibular joint (TMJ) disease is essentially a disorder of muscular inflammation and pain secondary to spasm. A strain or sprain may be precipitated by fatigue in the affected muscle secondary to a specific activity. The pain may be experienced as periocular if the trigger points in the temporalis, sternocleidomastoid, and trapezius muscles are affected, although pain also may be experienced as toothache, neck soreness, earache, neuralgia, and sinus discomfort because of the distribution of branches of the trigeminal valve, local muscle tenderness, jaw clicking and popping, and limitation in range of motion help to differentiate TMJ disease from other causes. Treatment is best managed by an oral and maxillofacial specialist.

Functional: Pain that has no pathophysiologic basis may have a supratentorial basis (e.g. somatization disorders, conversion hysteria, hypochondriasis, and schizophrenia). Secondary gain in litigious patients may be another basis for unexplained pain. After a thorough evaluation has failed to establish an organic basis, pain may be managed by analgesics, massage therapy, acupuncture, biofeedback training, and psychotherapy.

Other painful syndromes include photo-oculodynia, in which pain is elicited by light stimulus, and opthalmodynia periodica, in which sharp, intermittent, stabbing pains occur in trains of two or three paroxysms and do not occur again for weeks or months. Although there is no pathologic basis for either, the latter rarely may be associated with transient ischemia and may result from antiplatelet treatment.

Sažetak

Osećaj oftalmičkog/orbitalnog bola može da nastane direktnom ili indirektnom stimulacijom nervnih vlakana za bol i anatomskih struktura povezanih sa tim vlaknima. Bolna noksa može da pogodi trigeminalni živac i njegove grane, kao i druge živce: facijalni, glosofaringealni i vagus. Bol je nekad teško lokalizovati zbog toga što bolne senzacije konvergiraju od manjih grana ka glavnom narvnom stablu (na pr. zbog očnog bola koji se doživljava kao glavobolja ili akutni glaukom koji stvara oštar bol oka se često mešaju sa apscesom gornjeg molara). Osećaj bola može da bude oštar, ako ga prenose mala mijelinizovana A delta vlakna, ili tup, dugotrajni bol koji prenose mala nemijelinizovana C vlakna. Uopšteno, poreklo bola se klasifikuje kao da vodi od orbite, glave ili, ako se poreklo ne može odrediti, kao funkcionalan.

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