

MFs were found around the lining epithelium of 2/6 cases of mucous retention cysts, and in 3/12 cases of the adjacent to mucocoeles distended excretory ducts (Fig. 1C). Myoepithelial cells give to salivary glands parenchyma sufficient support but are absent from the lining epithelium of excretory ducts. Therefore, our results possibly indicate a muscular supportive role of MFs around the cystic wall of mucous retention cysts and distended excretory ducts.

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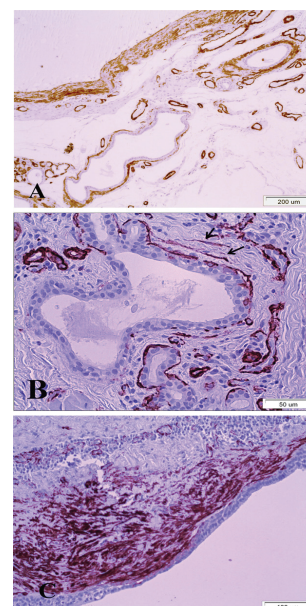
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**Figure 1:** A. Extravasation mucocoele where part of granulation tissue was substituted by dense connective tissue with large number of MFs. Also many MFs are presented around two distended excretory ducts adjacent to lesion. B. Small number of MFs around an excretory duct (arrows) in CS with severe degree of fibrosis. C. Large number of MFs is presented around the lining epithelium of mucous retention cyst (a-SMA Immunohistochemistry)



## Isolated sphenoiditis: presentation of 2 cases and review of the literature

Dear Editor,

Isolated Sphenoiditis (IS) is a relatively uncommon clinical entity (1-2,7% of all sinus infections). Due to important anatomic relations of the sphenoid sinus, the complications can be devastating. Predisposing factors for IS include forceful water entry into the nose during swimming or diving, allergic rhinitis, sinonasal polyps, bronchial asthma, septal deviation, middle/superior turbinate anomalies, radiotherapy, immunosuppression, diabetes mellitus and cocaine abuse. Among the pathogens that are involved in IS, the most common include *S.aureus*, *Streptococcus* species, *Peptostreptococcus*, *Fusobacterium*, *Klebsiella*, *P.aeruginosa*, *Aspergillus fumigatus* and *Aspergillus flavus*.

In our Department we have recently treated 2 patients with IS: a 45yo female and a 60yo female both complaining for severe occipital headache. Endoscopy revealed postnasal drip and purulent secretions from the left and right respectively sphenoid sinus ostium. CT scan showed complete opacification of the left sphenoid sinus in the first patient while a brain MRI (obtained earlier during a neurologic evaluation) revealed opacification of the right sphenoid sinus in the second patient. Initial conservative therapy (ceftriaxone 2g daily + clindamycin 600mg qid iv + xylometazoline 0,1% nasal spray) was unsuccessful.

Patients were finally admitted to surgery. Sphenoidotomy through endoscopic transnasal approach was performed. The postoperative period was uneventful and both patients were dismissed 48 hours after surgery. Patients remain free of symptoms and the new sinus ostia remain widely open.

The most common symptom on presentation is headache (50-60%)<sup>1,2</sup> with no characteristic pattern of distribution<sup>3</sup>. Patients may also experience ophthalmic involvement with VI or III cranial nerve paresis (diplopia in 40%) and blurred vision<sup>1</sup>. Classic symptoms of rhinosinusitis may also be present. Extension of the pathology causes orbital as well as intracranial complications.

Nasal endoscopy is useful for diagnosis. However, up to 60% of cases may have a normal endoscopic examination. Consequently, CT scan is the only modality that can consistently detect IS. The role of MRI is complementary. Initial management of IS is conservative (amoxicillin-clavulanic acid, and cephalosporins are typical choices). Response to conservative therapy, however, is often poor.

Surgical techniques developed in recent years include endoscopic transnasal approach, endoscopic trans-septal and endoscopic transpterygoid approach. The last two approaches are used for wide surgical exposure, in cases of suspected neoplasms.

The endoscopic transnasal approach is usually preferred. It can be performed with two approaches: (1) through the middle meatus following complete ethmoidectomy and (2) direct approach above the choanae. The transnasal technique using a direct approach is safest<sup>3</sup>. As an alternative to the use of an endoscope, successful use of a surgical microscope has also been reported<sup>4</sup>. Regardless of the selected surgical method, excellent control of the disease should be expected postoperatively.

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## Chronic inflammatory demyelinating polyradiculoneuropathy in childhood and response to IVIg

Dear Editor,

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an immune-mediated acquired polyneuropathy which is rare in childhood. The prevalence of patients aged under 19 years old with CIDP has been estimated at 0.48/100.000 and it is higher in male children than it is in female ones<sup>1</sup>. Children with CIDP present with subacute onset of symmetric proximal and distal weakness that progress over at least 2 months. The clinical criteria for the diagnosis of CIDP include: a) progressive or relapsing motor and sensory dysfunction of more than one limb and b) hyporeflexia or areflexia that usually involves all four limbs. Moreover, children with CIDP present with elevated cerebrospinal fluid protein, diminished motor and sensory conduction velocities during electrophysiological studies and also typical demyelinating deterioration in possible sural nerve biopsy<sup>2</sup>. The first-line therapy is IVIg, while the alternative choices include corticosteroids, haemodialysis or immunomodulatory agents<sup>2</sup>.

This is the case of a 3 year old boy referred in our department afebrile and in good condition. Parents reported weakness after 500 metres of walking, frequent falls, weakness when climbing a ladder. Neurological examination revealed hypotonia of lower limbs and diminished tendon reflexes. Blood tests and biochemical examinations including CPK (Creatine phosphokinase), metabolic screening (plasma ammonia, lactic acid, plasma and urine aminoacids, homocysteine, phytanic acid), thyroid gland check and gene screening for spinal muscular atrophy were normal, while in the cerebrospinal fluid (CSF) showed raised protein (72mg/dl, normal limits <45mg/dl) and normal oligoclonal bands. Anti-GM1 IgM (Anti-ganglioside M1) serum antibodies were detected, while the rest antibodies against gangliosides (Anti GD1b, Anti-GQ1b, Anti-MAG) and rest immunologic tests (ANA, anti-DNA, ENA, ANCA, ASMA, RF, anti-AChR, anti-MuSK) were negative. Clinical examination (absence of tendon reflexes) and raised CSF protein levels raised the suspicion of a possible demyelinating polyneuropathy. Following spinal and brain Magnetic Resonance Imaging (MRI) were normal and electrophysiologic studies showed distal demyelinating polyneuropathy and confirmed the diagnosis (diminished sensory conduction velocities and prolonged distal latencies of median and ulnar nerves, conduction block in all affected nerves of upper and lower limbs, absent f wave latencies in right ulnar and left peroneal nerves). Intravenous human immunoglobulin (IVIg) in dose 0,4 mg/kg/D for 5 days was initially given, followed by tapering monthly doses of 1 g/kg for 10 months. The patient had a very good response to treatment with IVIg, with acute improvement of muscle weakness within 4 days after the administration. The clinical improvement after each administration of IVIg lasted about 4-6 weeks. The child also started physiotherapy sessions and 10 months later remains completely asymptomatic.

Children with CIDP, as in our case, have a more favorable outcome than adults, with good response to IVIg or corticosteroids<sup>2</sup>. In this paper we want to mention the clinical and laboratory profile of CIDP, and also the necessity of appropriate diagnosis and treatment in a timely fashion, so patients can have a favorable outcome.

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ANA: Anti-nuclear antibody, **anti-DNA**: Antibody to Deoxyribonucleic Acid, ENA: Antibody to Extractable Nuclear Antigen, ANCA: Anti-neutrophil Cytoplasmic Antibody, ASMA: Antismooth Muscle Antibody, RF: Rheumatoid Factor, **anti-AChR**: Anti-acetylcholine receptor antibodies, **anti-MuSK**: Antibodies to muscle-specific receptor tyrosine kinase