CLINICAL PHOTOGRAPHS

Hereditary hemorrhagic telangiectasias

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant disorder characterized by recurrent epistaxis, cutaneous telangiectasia, and visceral arteriovenous malformations that affect many organs, including the lungs, gastrointestinal tract, liver and brain. HHT affects varied racial and ethnic groups and occurs in a wide geographic distribution. Men and women are affected equally. Onset of symptoms may be delayed until the fourth decade of life (approximately 90% of patients manifest by age 40 y) or later decades. The HHT diagnosis is classified as definite if 3 criteria are present, possible or suspected if 2 criteria are present, and unlikely if fewer than 2 criteria are present: recurrent epistaxis, telangiectasias - multiple at characteristic sites (lips, oral cavity, fingers, nose), family history, visceral lesions (gastrointestinal telangiectasia with or without bleeding). Laboratory examinations can reveal a low value of hemoglobin, because of chronic bleeding, platelets may have normal value while coagulation is normal. Imaging (CT, MRI) may reveal the existence of vascular-nerve abnormalities. The surgical treatment consists in electrocoagulation or photocoagulation of the telangiectases from the nasal mucos, or arteriovenous aneurysm embolization.

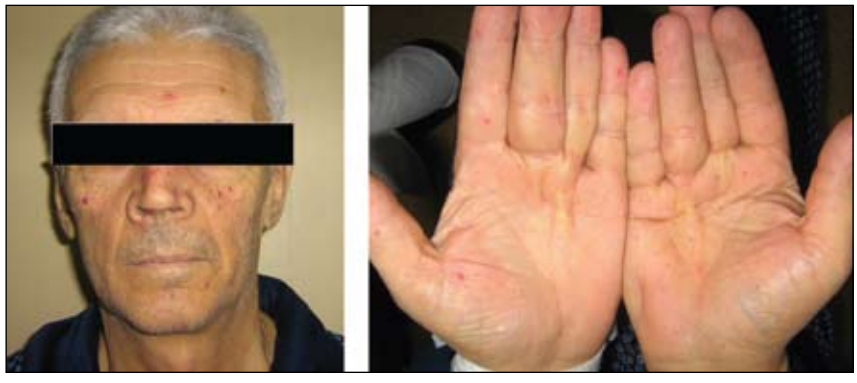

Figure 1 a, b Telangiectasias on the face skin and hands

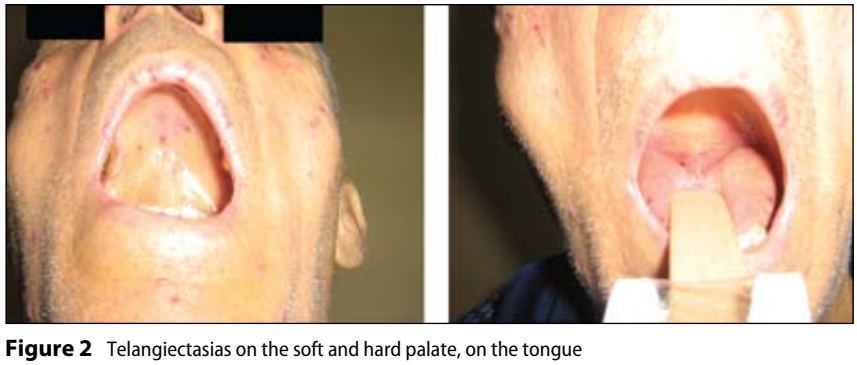

Figure 2 Telangiectasias on the soft and hard palate, on the tongue

Nasal fibrosarcoma

Fibrosarcoma arising in the sinonasal cavities are very rare. Fibrosarcoma is a tumor of mesenchymal cell origin that is composed of malignant fibroblasts in a collagen background. The tissue of origin seems to be the periosteum rather than mucosal connective tissue. Fibrosarcoma, like other soft-tissue sarcomas, has no definite cause. Current research indicates that many sarcomas are associated with genetic mutations. The most common genetic defects include allele loss, point mutations, and chromosome translocations. Also, their etiology could involve previous trauma, nasal polyposis and a history of radiation therapy. Fibrosarcoma is a malignant tumor with a high cellularity, the tumoral cells are spindly, uniforms, without pleomorphism, with a moderated to high mitotic rate. The most common presentation for sinonasal fibrosarcomas includes epistaxis, nasal obstruction, recurrent sinusitis, cranial neuropathy, sinus pain, facial paresthesia, proptosis, diplopia.

Because of the rarity of these tumors no clear guidelines exist for their therapy. Regarding the treatment, depending on the type and extent of the tumor, surgical, radiotherapeutic measures are appropriate. Inclusion of neoadjuvant chemotherapy in modern therapy regimens may facilitate resection of unresectable tu-

mors and improve the outcome of these patients.

Young 29 years old patient with history of nose and sinuses surgical procedures - septoplasty (2007), septal button placement (2008), nasal polypectomy (2008), excision of a mesenchymal tumor of the right nasal fossa (2008) - presented for nasal obstruction.

Anterior rhinoscopy and nasal endoscopy revealed a tumor that involves 2/3 of both nasal cavities and affects the superior part of the septum by destroying it. Both middle turbinates were laterally moved and an important pressure was made on lateral walls of the nose.

The CT and MRI scans showed a voluminous tumor that involved nasal cavities, ethmoid, sphenoid, frontal sinuses with invasion of medial walls of maxillary sinuses and orbital cavities and intracranial extracerebral extension.

Surgical approach was endoscopic endonasal with a piecemeal resection. Histopathologic examination and imunohistochemistry test establish the diagnosis of fibrosarcoma. Postoperative the pacient received radiotherapy.The endoscopic and imaging (CT scan) reevaluation at 2, 3 and 5 months shows no recurrence.

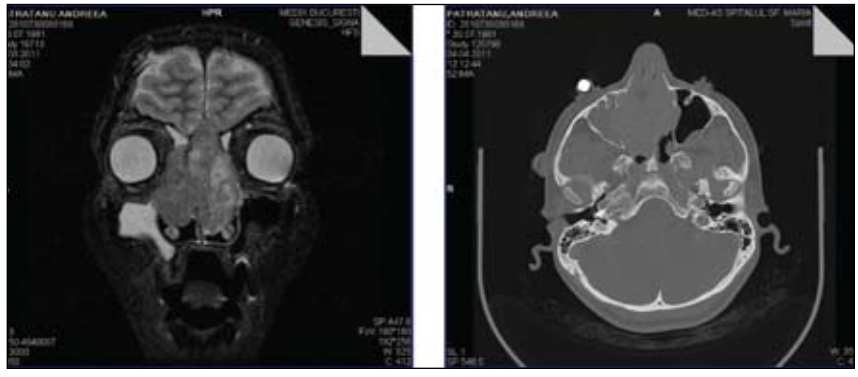

Figure 1 Preoperatively IRM scan (coronal slice) and CT scan (axial slice)

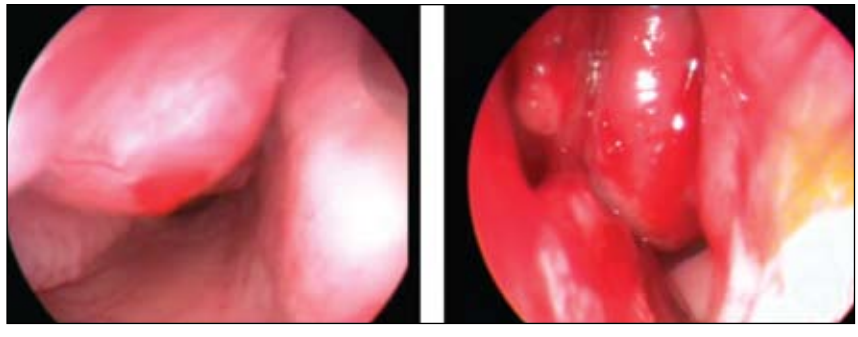

Figure 2 Intraoperatory view

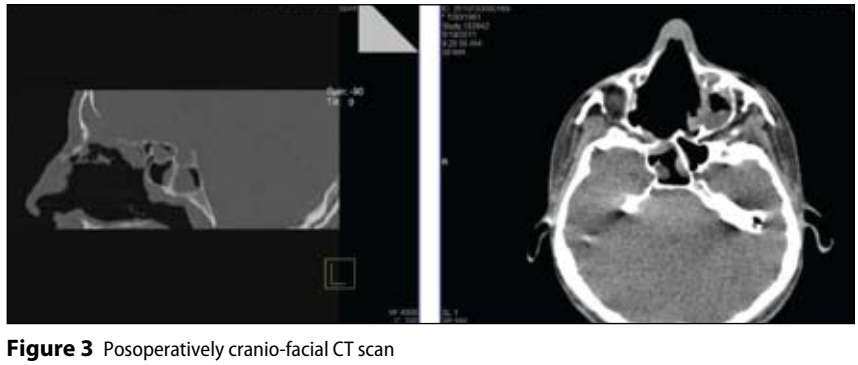

Figure 3 Posoperatively cranio-facial CT scan