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Case Report
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Syphilitic Aortitis with Thrombosis of Internal Carotid Artery Sinistra-Case Report

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Abstract

Otolaryngologists rarely come across with syphilis cases, and it hampers diagnosing. We mean patients with primary, secondary and tertiary syphilis. Patients with primary laryngeal syphilis have larynx most frequently attacked (erythematous, erosive and ulcerative chancre), nasal cavity is least attacked. With secondary syphilis local occurrences in larynx or pharynx are accompanied by rash on skin of trunk and extremities. Pharyngoscopy of secondary laryngeal syphilis is characterized by diffuse swelling associated with copper-red hyperemic pillars of thefauces, mucous membrane of soft and hard palate. Secondary stage in pharynx occurs in the form of erythema, simulating catarrhal laryngitis driven by involvement of plicavocalis, arytenoid cartilage and epiglottis in the process. Papulae are likely to appear with localization in different parts of larynx and pharynx. Tertiary syphilis of ENT organs can be localized in any part of upper air passages (nose, pharynx, palate, larynx) and manifests itself as circumscribed gummous tumor, which can ulcerate [1]. According to WHO classification of syphilis, there are congenital, early (primary and secondary) and late (tertiary – cardiovascular, neurosyphilis and lesion of other organs) [2].

Over the past 80-120 years there is a conspicuous tendency in the change of localization of late forms of syphilis. There is a drastic decrease of frequency of registration skin and bone manifestations (from 39 to 2% according to Fournier and M.V. Milich), escalation of registration of cardio-vascular registration of syphilitic cases from 1 to35% , decrease of registration of visceral syphilis (from 27 to 0,5%) and relatively high ratios of neurosyphilis (from 33 to 62%). Neurosyphilis patients have been redistributed within the group. Decreased frequency of parenchymatous form (amyelotrophy, progressive paralysis), mitigation of the clinics as well as increased number of patients with mesenchymal form is driven by meningovascular and vascular syphilis of cerebrum and spine [3,4]. This case is presented to emphasize possible development of syphilitic arteritis of internal carotid artery. Herewith, we present the case of the patient, who appeared in our division with suspicion of nasopharyngeal tumor.

Abbreviations: MRT: Magnetic Resonance Tomography; CT: Computed Tomography; TPIT: Treponemapallidum Immobilization Test; IT: Immunofluorescence Test; ELISA: Enzyme-Linked Immunoassay

Case Report

Patient K was born in 1965, addressed to the division of inflammatory diseases of State Institution «O.S. Kolomiychenko Institute of otolaryngology of NAMS of Ukraine", having been referred by ENT doctor of a local hospital. Patient complained of periodic sharp shooting facial pain on the left side and diminished hearing in the left ear over the last 2 months. He had an acute nasal hemorrhage from left nostrill a week before his visit. He had to address to local district of medical community. They helped him stop the hemorrhage. Then the patient was examined in the therapeutics division. The patient was diagnosed with diabetes mellitus, anisocoriasinistra and neuralgia of trigeminussinistra. The patient was asked to go through additional examination and treatment at neurological division of Zhytomyr regional hospital, where he took magnetic resonance tomography (MRT) and computed tomography (CT) of temporal bones and cerebrum. He was diagnosed with nasopharyngeal tumor. Secondary neurology of trigeminus of rami I-II with severe pain syndrome. Diabetes mellitus type 2, non-insulin-dependent diabetes mellitus, moderately severe, sub compensation state.

i. **Objective Examination:** General state is satisfactory, no sleep and appetite disorders.

Blood pressure: 140/85 mm Hg., regular pulse: 82 bpm, body temperature: 36,7 °C. Anisocoria is observed (left pupil contracted).

ii. **Otoscopically:** Retraction of left tympanic membrane, diffuse light reflex.

iii. **Right Tympanic Membrane:** No peculiarities. Rhinoscopy – deviated septum to the left side (along the crest). Endoscopy in nasopharynx showed bulging mucous membrane, mostly from left side. Mucosa was pale pink, without pathological inclusions or deposits, the structure

looked like adenoid vegetations. Pharyngoscopy showed no peculiarities.

iv. CT Made in the Place of Residence Showed: Tumor in nasopharynx with the signs of infiltrating growth. Chronic left medial otitis, mastoiditis (Figures 1A & 1B).

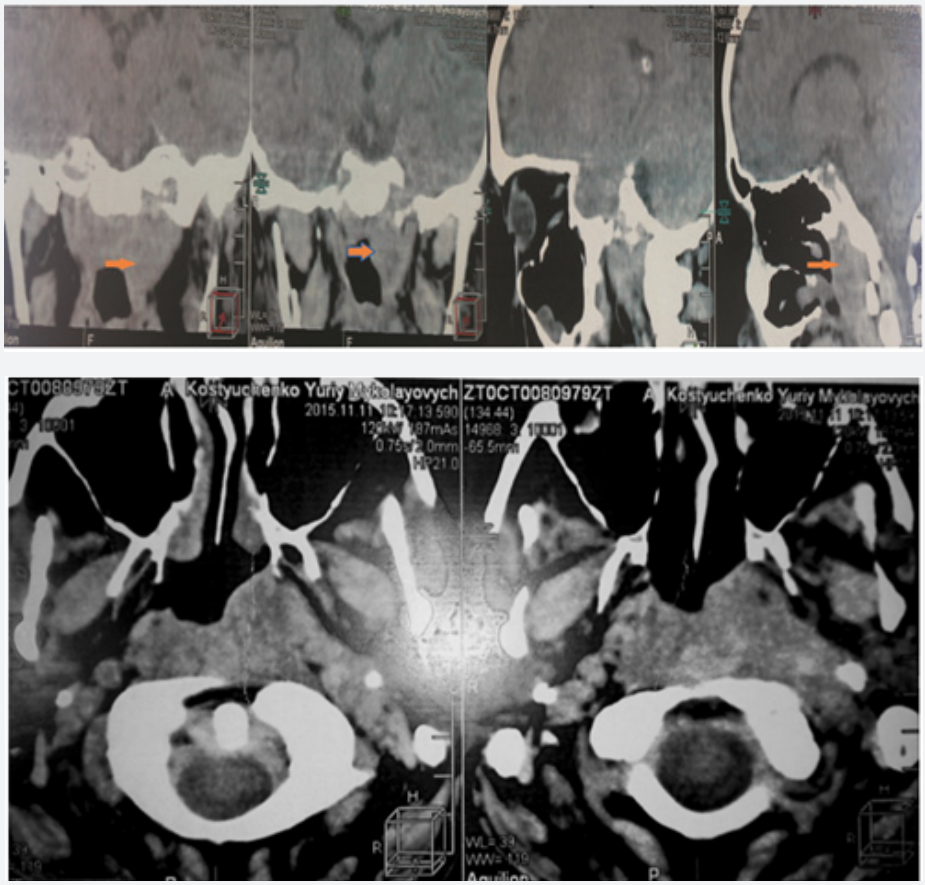

Figure 1: CT of scull base of 11.11.15.

v. MRT Findings: Massive cerebral microangiopathy, dyscirculatory encephalopathy Consequences of sinistraloto-mastoiditis. Retention cysts of the maxillary antrum (Figure 2). Findings of Audiometry, performed at local hospital on 21.10.15: bilateral sensorineural hearing loss I stage, tubootitis sinistra.

To verify expansion of the tumor, its blood supply and the safest point for biopsy, the patient was asked to take head CT with contrast material. CT as of 17.11.15 showed: tumor of irregular shape, sized circa 73×33×40 mm of inhomogeneous density, with necrotic patch, with heterogeneous contrast enhancement, with the signs of destruction of skull base bones in nasopharynx along posterior sinistral wall, expanded to its anchoring. Left internal carotid artery breaks in full- thickness of tumor, starting at 10 mm from bifurcation level to the siphon and internal jugular vein (invasion signs). There are numerous posterior lymph nodes sized 10×6 mm from both sides.

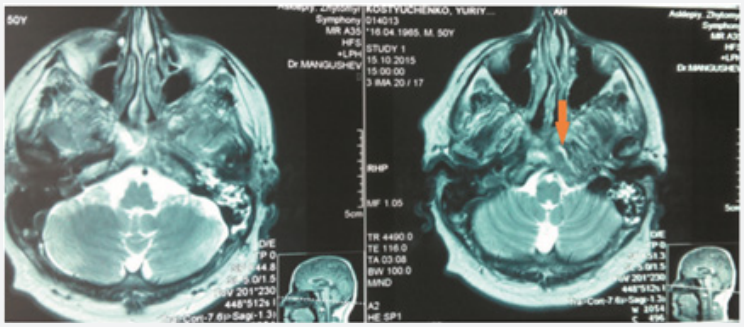

Figure 2: MRT of brain 15.10.15. Findings of Audiometry, performed at local hospital on 21.10.15: bilateral sensorineural hearing loss I stage, tubootitis sinistra.

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vi. Medical Report: CT- there is a picture of tumor in nasopharynxsinistra with destruction in bones of cranial base – cuneate and occipital, invasion of internal carotid artery and sinistral internal jugular vein. Cervical lymphadenopathy is most likely of secondary aplastic genesis (Figures 3A-3C). Multidetector computed tomography of head and soft cervical tissues with IV contrast – Ultravist – 370,0 – 100,0 ml). Since internal carotid artery and internal jugular vein broke off in tumor thickness, it was necessary to assess blood supply to take a safe biopsy. It was decided for patient to undergo direct selective angiography of this section.

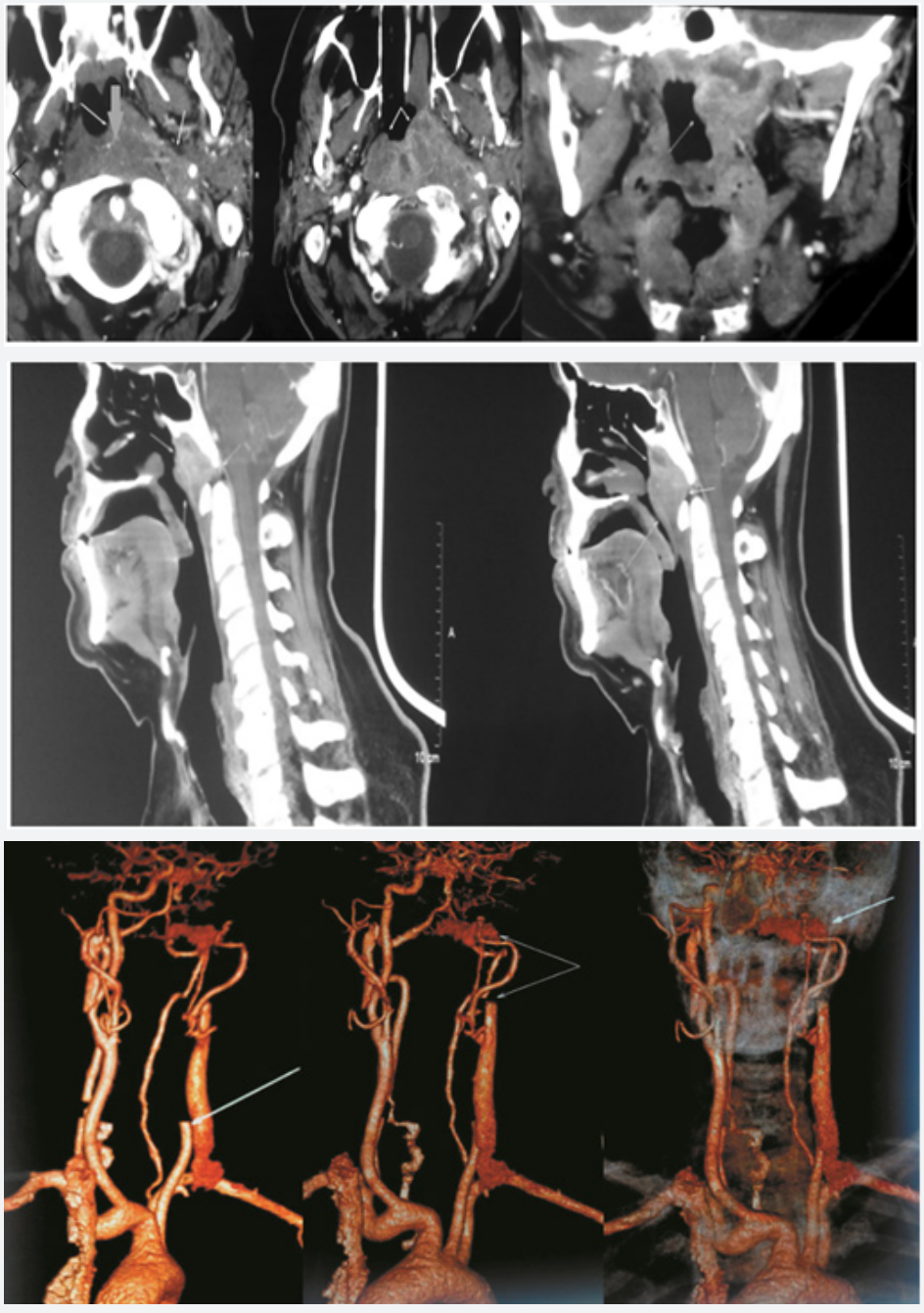

Figure 3: Multidetector computed tomography of head and soft cervical tissues with IV contrast – Ultravist – 370,0 – 100,0 ml).

vii. Angiography Result: Defined thrombosis of left internal carotid artery, filling of left carotid arterial system by anterior cerebral artery, anterior communicating artery; external carotid artery is without peculiarities. Tumor is not filled with contrast. Upon results of direct selective angiography, the patient was taken to the operation room for biopsy innasopharynx under general anesthesia and endoscopic control 0°. A tissue similar to adenoid mass was found. Arch incision in the part of the largest bulging was made with sickle knife under endoscopic control. There was a scarce purulent discharge from the incision.

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With aspiration of purulent discharge, exsufflator detected whitish conglomerate in the thickness of the neoformation. The latter was removed with Blakesleyforceps. Conglomerate is sized 80×30×20mm, of green and yellow color with odor, dense consistency (Figure 4).

Conglomerate and sections of neoformation were taken for pathohistological study. The results show combination of chronic and acute inflammation with numerous plasmatic cells, necrotic patch and fibrosis of connective tissue, thrombosing of vessels. Lues and systemic lesion of vessels are to be excluded.

Having received the results of pathohistological study of patient, anamnesis vitae was supplemented. The patient «confessed» that he actually had syphilis 15 years ago and underwent the treatment in local hospital. Considering the data of anamnesis, clinics and pathohistological study, the patient was asked to undergo serological blood test, treponema - Wasserman reaction (qualitative and quantitative methods), treponemapallidum immobilization test (TPIT), immunofluorescence test (IFT), immunofluorescence test -200 (IT-200), enzyme-linked immunoassay (ELISA), Enzyme multiplied immunoassay (EMIT).

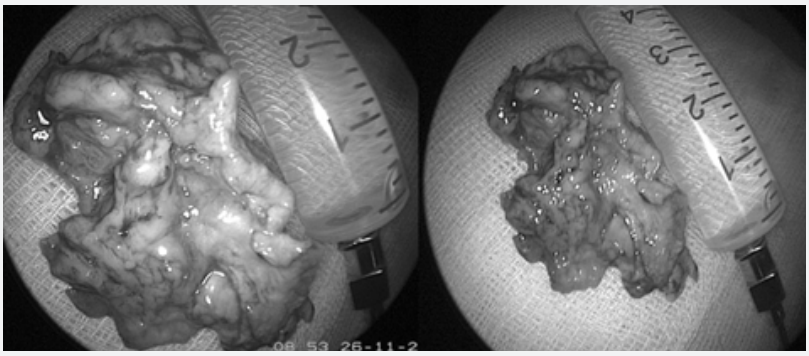

Figure 4: Removed conglomerate from nasopharhynx.

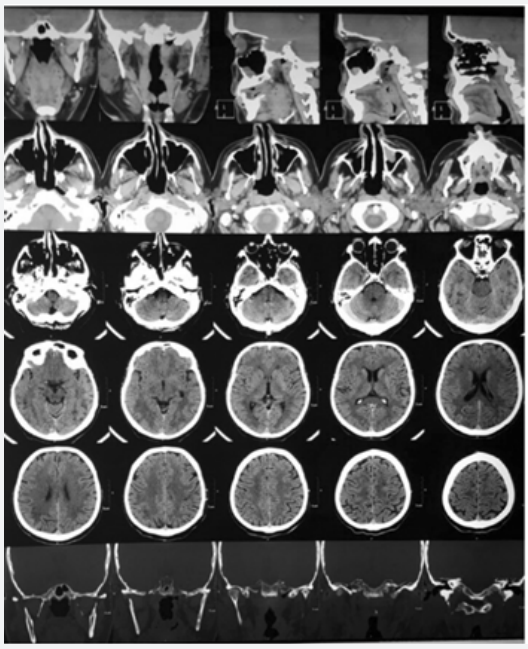

Figure 5: CT images of scull base after treatment.

Result

Wasserman reaction, TPIT - negative, IFT-200 and EMIT are weakly positive, IFT – positive. Patient was consulted by dermatovenerologist and diagnosed with tertiary syphilis, gummous form (nasopharyngeal gumma) with the expansion and to petrous apex and destruction in bones of cranial base – cuneate and occipital, invasion of internal carotid artery and left internal jugular vein. Patient was prescribed with G, 2. million units IM trice a day for 16 days, 3 courses, with the interval of one month. Upon termination of the first course, the patient felt better, had no headache. His hearing on left ear and nasal breathing got better.

Discussion

Vascular syphilis of head and neck and late diffuse meningovascular syphilis are currently major forms of late

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neurosyphilis. At present vascular manifestations are observed in 70% [4]. Meningovascular syphilis makes circa 3% of total diagnosed syphilis cases and about 10% of neurosyphilis [5]. In case of meningovascular syphilis, meninx and brain substance are not involved. Thus, the composition of cerebrospinal fluid remains normal. The disease is mostly observed in male group aged 30-50 years. It is characterized by development of specific infiltrate in the cerebral vessels, which causes thrombosis or stroke [6]. Vascular syphilis is resulted in infection-associated inflammatory arteriopathy of cerebral vessels, vessels of spinal cord and neck. Histopathological picture is morphologically similar to autoimmune arteritis of central nervous system (SNS). Diffuse thickening and lymphocytic infiltration of meninx vasculosa of meninxvasculosa and perivascular space are generally observed. The most common form of syphilitic arteritis is Heubnerarteritis, where large and medium vessels are injured. In case of syphilis, injury of small vessels is known as Nissl-Alzheimer arteritis.

Finally, in both cases [6,7] obliteration and much rare aneurysmal ectasia take place. The diagnosis is complicated due to the fact that in 60-70 % of cases serological reactions (Wasserman, Kan, Zax-Vitebsky) are negative. Depending on localization of injured vessels, neurological symptomatology may differ. The following clinic symptoms are important in diagnosing the disease: papillary disorders (miosis, mydriasis, anisocoria, pathognomonic symptom of Argyll Robertson, in particular), drop out of knee and ankle jerk reflex Achilles Reflex, Romberg ataxia , bone and air dissociation , trophism disorder , cold hyperesthesia in the back , pain hypesthesia in thoracic zone (Hitzig zone). Argyll Robertson syndrome remains even after efficient treatment. In most cases, described in the manuals, patients with syphilitic thrombosis of internal carotid artery are hospitalized with the signs of ischemic stroke, in indolent progression, with likely remission periods. Late vascular cerebral syphilis can be often combined with other forms of neurosyphilis, viz. amyelotrophy and visceral syphilis. Likely combination of atherosclerosis and syphilis of cerebral blood vessels should be noted. The senior the patient, the most frequently this pathology occurs [8].

Analyzing syphilitic manifestations of head and neck vessels, including internal carotid artery (Holland et al, Brightbill et al, Tien et al, Asdaghi et al, Umashankar et al.) we can observe that internal artery is rarely involved in the process, mostly jointly with medium and posterior cerebral artery. Clinical data are associated with development of necrosis of cortex or subcortex structures of brain. Keen interest and novelty displayed in this case is accounted for complete exclusion of left internal carotid artery and the fact that there are no clinic features connected with its obliteration. Minimal clinic symptoms can be connected with formation of gummous process in nasopharynx, and subsequently, thrombosis of internal carotid artery. Otherwise, formation of syphilitic arteritis with thrombosis of internal carotid artery can be primary, and with time specific inflammatory process can be restricted; and gumma can be developed further.

Conclusion

Finally, we can infer that vascular syphilis is extremely difficult to diagnose, as most patients have asymptomatic course, absconding as atherosclerotic vascular disease with patients of senior age. Even specific serological reactions are frequently negative. Patients often do not focus on syphilis, which either remained without treatment or adequate treatment, or conceal this fact. Thus, a strong psychological rapport with patient and comprehensive anamnesis are of paramount importance [9].

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