PATHOLOGIES OF PETROUS APEX

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The review of papers, focused on studying various neoplasms, diagnosis, selection of surgical approach, complications, and recurrence rates of the petrous apex lesions, that have been published in 2008–2022, is provided. Effusion, mucocele, cholesterol granuloma, cholesteatoma are the most common benign lesions of the petrous apex. Such surgical approaches as translabyrinthine, transcochlear, endoscopic endonasal approach and the middle cranial fossa approach are most often used during treatment. The middle cranial fossa approach, infracochlear approach and endoscopic transnasal approach are recommended for patients with preserved hearing. In case of disseminated lesions, when the carotid artery and the jugular bulb should be additionally controlled, transcochlear and translabyrinthine surgical corridors could be used.

Keywords: petrous apex, neoplasm, middle cranial fossa, surgical approach, skull base, internal carotid artery, facial nerve, liquorrhea, complications, cholesteatoma, cholesterol granuloma, effusion, diagnosis

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ПАТОЛОГИЯ ВЕРХУШКИ ПИРАМИДЫ ВИСОЧНОЙ КОСТИ

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Представлен обзор публикаций, посвященных изучению различных новообразований, диагностики, выбора хирургического доступа, осложнений и частоты рецидивирования патологии верхушки пирамиды височной кости за период с 2008 по 2022 г. Наиболее распространенной доброкачественной патологией верхушки пирамиды височной кости являются выпот, мукоцеле, холестериновая гранулема, холестеатома. При лечении чаще всего используют такие хирургические доступы, как транслабиринтный, транскохлеарный, эндоскопический эндоназальный и доступ через среднюю черепную ямку. Для пациентов с сохранной слуховой функцией рекомендовано использование доступов через среднюю черепную ямку, инфракохлеарного, либо эндоскопического трансназального доступов. В случае распространенных патологических процессов, когда необходим дополнительный контроль сонной артерии и луковицы яремной вены, могут быть использованы транскохлеарный либо транслабиринтный хирургические коридоры.

Ключевые слова: верхушка пирамиды височной кости, новообразование, средняя черепная ямка, хирургический доступ, основание черепа, внутренняя сонная артерия, лицевой нерв, ликворея, осложнения, холестеатома, холестериновая гранулема, выпот, диагностика.

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The petrous apex, located in the petrous part of the temporal bone, is an anatomical area difficult for surgical treatment, since it lays adjacent to vital anatomical structures (second segment of the internal carotid artery (ICA), cavernous sinus, cranial nerves V and VI, temporal lobe of the brain, subarachnoid cistern).

Various pathologies are found at the site, among which space-occupying lesions, that require only surgical treatment, are the most common, and selection of optimal approach remains the main problem in such cases. Extradural and intradural lesions of the petrous apex are distinguished based on etiology. The most common extradural lesions are cysts (asymmetric effusion), which are more prevalent than cholesterol granulomas, cholesteatomas, chondromas, mucocele. Intradural lesions include meningioma and schwannoma [1, 2]

The petrous apex lesions are usually incidental findings, they do not produce any clear symptoms given the small mass size and petrous pyramid limited with bony walls. This makes them difficult to diagnose. Disruption of bony edges of the petrous apex and compression of important anatomical structures produce a vivid clinical picture (pain on one side of the face (trigeminal nerve (cranial nerve V)) or headache, diplopia (abducens nerve (cranial nerve VI)), weakness of facial muscles (facial nerve (cranial nerve VII)), hearing loss and vestibular disorders (vestibulocochlear nerve (cranial nerve VIII)), Gradenigo's syndrome) that motivates the patient to seek urgent medical care [3, 4]. The diversity of symptoms results from the petrous apex position on the boundary between the otorhinolaryngological and neurosurgical practices. Surgical treatment of patients with the pathologies of the petrous apex is difficult due to the petrous apex medial position at the base of the skull, location in the close proximity to vital anatomical structures, and therefore high risk of injury to these natomical structures during surgery. Surgical techniques are limited to the use of microscopic, endoscopic or combined methods involving creation of the anterior (from the upper surface of the pyramid or transnasal), lateral (translabyrinthine, transcochlear, infracochlear approach) or posterior (retrosigmoid approach in case of intradural lesion) surgical corridor.

Each of these approaches has certain advantages and disadvantages. To date, no standard surgical algorithm has been developed. Some patients do not receive adequate treatment due to the surgeons' (otosurgeons'/neurosurgeons') lack of experience with this zone; there are almost no publications on this topic in Russian journals. In case of suspected benign lesion the need to select a gentle treatment strategy involving maximum preservation of adjacent anatomical structures is noted.

The review describes the most common petrous apex lesion, surgical approaches and the petrous apex anatomy.

Epidemiology and pathophysiology

Asymmetric pneumatization and effusion in the cells of the petrous pyramid are the most common manifestations of the petrous apex lesions [5]. The estimated incidence of the petrous apex granuloma is 0.6 cases per 1 million population. Cholesterol granuloma is 10 times more prevalent than the petrous apex cholesteatoma and 40 times more prevalent than mucocele [5].

There are two theories about the cholesterol granuloma formation. The obstructive vacuum theory is based on the obstruction of air space resulting in gas absorption an resorption, vacuum, mucosal hemorrhage, inflammation, and degradation of red blood cells followed by formation of cholesterol crystals. During the cholesterol crystal formation, the inflammatory cascade is triggered by bone resorption and inflammatory body reaction [6]. The theory of the spongy bone involvement in based on the development of abnormal link between the air cell system mucosa during intense pneumatization of the petrous apex with subsequent gradual replacement with spongy bone accompanied with hemorrhage into the petrous apex air cells [6]. Red blood cells decay to the form cholesterol crystals, thus inducing the inflammatory cascade [6].

The hypothesis has been proposed that cholesterol granulomas are formed due to eustachian tube dysfunction, negative pressure not only in the tympanic cavity, but also in mastoid cells and cells of the petrous pyramid, which communicate with the tympanic cavity, most often indirectly [7]. The resulting negative pressure leads to the leakage of fluid (plasma) from the submucous blood vessels through the mucous membrane of the tympanic cavity, mastoid cells and cells of the petrous pyramid. In some cases, blood vessel rupture and hemorrhage between the mucous membrane and bony wall occur in certain areas of the mucous membrane [7]. In the conditions of oxygen deficit, hemolysis of red blood cells (hemoglobin) occurs that also results in the formation of cholesterol crystals from cell membranes, causing the giant cell reaction, which in turn leads to the cyst formation [7]. When the bone surface becomes deficient, with coaptation of richly vascular mucosal air cell lining, accumulation of blood leads to abnormal blood outflow pathways, and increased accumulation of blood enables progressive cyst expansion [7]. As the cyst expands, bone erosion increases the area of exposed bone surface along the cyst wall [7]. Based on the above theory, is was shown that six patients with cholesterol granuloma out of 13 definitely had a gap between the bony wall and the mucous membrane of the petrous apex air cells, while the control patients with pneumatized apices showed no signs of such gap [7]. This theory was confirmed by additional hystological assessment of the temporal bone.

Cholesteatoma (epidermoid cyst) consists of the epithelial torus structure, subepithelial fibrous tissue, and keratinized desquamated debris. It is believed that the remaining epidermis is formed in close vicinity to the foramen lacerum during embryonic development [6] or due to migration of ectoderm of the external auditory canal [6]. Expansion of cholesteatoma may result in bone erosion due to osteolytic enzymes of epithelium and subepithelial fibrous tissue. Epidermoid and dermoid cysts can be distinguished by the presence of skin appendages (for example, sweat glands). Mucocele, which is formed due to obstructed drainage from the excessively pneumatized petrous apex, can also cause the development of cystic mass [6].

Diagnosis

Primary lesions of the petrous apex are usually diagnosed at late stages since such lesions are rare and asymptomatic at early stages. Defects of the petrous apex and around it could be caused by cholesteatoma, meningioma, schwannoma, giant cell tumor, cholesterol granuloma, ICA aneurysm, metastases or mucocele. The clinical features result from the compression of cranial nerves IV, V and VI. Larger lesions may affect the cranial nerve complexes VII–VIII. Erosion in the dura mater may result in otoliquorrhea [8]. Masses in the petrous apex are revealed mostly by CT and MRI. These diagnostic methods are also used to define the postresection cavity size and exclude postoperative complications.

Surgical treatment

Surgical treatment of the petrous apex lesions is recommended to patients with such symptoms, as hearing loss and vestibular disorder, i.e. when the lesion spreads beyond the petrous apex and extends to the labyrinth.

The main factor that should be analyzed prior to surgery in case of the petrous apex lesion is the lesion localization: extradural (cholesterol granuloma, clival chordoma or chondrosarcoma) or intradural (meningioma, epidermoid and dermoid tumors). This is the only factor that provides a key to understanding of both etiopathogenesis and the question of which neural and vascular components might be involved in the process [9]. Furthermore, surgical treatment is selected based on the assessment of dysfunction of the facial and vestibulocochlear nerves [9, 10].

Several surgical approaches to the petrous apex have been described, which could be classified as transtemporal/transnasal or hearing-preserving/non-hearing-preserving [5]. Surgical treatment includes mass excision or marsupialization (minor surgical procedure to drain out cysts or granulomas). Excision (for example, of cholesterol granuloma) usually involves wide exposure of the lesion and often requires craniotomy with appropriate temporal lobe retraction. The *advantage* of excision lies in the chance to reduce the number of relapses, and *disadvantages* lie in the need for brain retraction (which may result in encephalomalacia and sometimes in seizures) and the increased risk of postoperative intracranial hemorrhage [5–7]. Liquorrhea also contributes to the risk of intracranial surgery, especially in patients with cholesterol granulomas, who usually demonstrate hyperpneumatization of the temporal bone [11].

A major shortcoming of marsupialization is the relapse of symptoms due to scarring that blocks the surgical drain [5].

A clinical case is reported of the 24-year-old patient with cholesterol granuloma complicated by Gradenigo's syndrome, who initially presented with nausea, fever, photophobia, left retro-orbital pain, and headache [11]. At first, the following diagnosis was established: bacterial meningitis based on the data of lumbar puncture due to detection of *H. influenzae* in cerebrospinal fluid (CSF); the patient received a course

of antibiotic therapy. After a few days, diplopia emerged in addition to the listed above symptoms.

Brain MRI revealed an expansile erosive cholesterol granuloma, which was removed during the combined operation performed by neurosurgeon and otorhinolaryngologist with endoscopic transsphenoidal drainage. Following evacuation of the cyst contents, the left petrous apex defect was marsupialized using the right middle turbinate mucosal graft, harvested earlier in the appropriate procedure. Histopathology revealed xanthogranulomatous inflammation consistent with the radiologically suspected cholesterol granuloma [11].

Such atypical manifestation of Gradenigo's syndrome with the rare complication of meningitis associated with granuloma of the petrous apex demonstrates the importance of early detection, mandatory use of radiological assessment methods (MRI), and consideration of the possibility of performing surgery in patients with pre-existing lesions of the petrous apex, who are potentially at higher risk of dangerous complications. Thus, surgical approach and operation procedure are selected based on the clinical manifestations, lesion, space-occupying lesion site, position of blood vessels, and surgeon's experience. Anterior, lateral (posterior) and anterolateral (transpetrosal) approaches to the petrous apex are distinguished. Anterior approaches include the following: medial transsphenoidal approach; medial transsphenoidal approach with lateralization of the ICA; sphenoidal approach in the lower petrous part of the temporal bone; endoscopic approach through the foramen lacerum; endoscopic anterior resection of the petrous apex. Lateral (posterior) approaches include: translabyrinthine, transcochlear, transcanal infracochlear, and the middle cranial fossa approaches. Anterolateral (transpetrosal) approaches are as follows: open anterior resection of the petrous apex.

Translabyrinthine approach

It is considered a standard otosurgical approach that provides the widest lateral corridor to the petrous apex with the best control over the the entire length of the facial nerve. However, this approach entails removal of semicircular canals, vestibule, and identification of internal auditory canal. The approach provides limited access to the petrous apex. The risks of this surgical procedure include the development of liquorrhea, facial nerve injury, vestibular disorders, and the loss of residual hearing.

Infracochlear or supracochlear approach

It is indicated for lesions located under the internal auditory canal between the jugular bulb, ICA and cochlea [8–10,12].

When using this approach, the bony wall of the lowest basal cochlea curl, jugular bulb and petrous segment of the carotid artery are the key targets that are exposed and identified within the medial wall of meso-hypotympanum. Sometimes, larger and more lateral cholesterol granulomas are found in the lateral part of the infralabyrinthine tract, which can easily expand into the middle ear. The medial cholesterol granulomas require more extensive resection that could be sometimes difficult due to the presence of jugular bulb or the petrous segment of the carotid artery located posterior at this site. The risk of liquorrhea increases when the resection angle points to the posterior cranial fossa (PCF) and cochlear aqueduct. Hearing loss and vertigo may develop in case of injury to the cochlear aqueduct. Injury to the carotid artery is rare, however, it requires emergency angiographic intervention [12].

Transotic approach

It provides access to entire petrous apex. With this approach, the same stages are used as in the translabyrinthine aproach, with the mandatory closure of the external auditory canal and removal of the posterior wall of the carotid canal and cochlea. The facial nerve is preserved in the canal, unlike the transcochlear approach that involves the backward shift of the entire intratemporal part of the nerve. Transotic approach has the same risks, as the translabyrinthine aproach. Injury to the carotid artery and iatrogenic cholesteatoma formation due to incomplete removal of skin from the external auditory canal are also possible. Transotic and transcochlear approaches usually require eustachian tube obliteration, since liquorrhea is often observed during cochleectomy. These approaches make it possible to perform marsupialization, and in some cases complete excision of the neoplasm.

Transcanal transpromontorial approach

It is an improved method for endoscopic transcanal transpromontorial approach developed for patients with hearing loss or no hearing [9,10,12] that is indicated for removal of lesions located in the internal auditory canal and extended to the petrous apex, more medial to the ICA [8, 9]. This is the access between the tympanic and mastoid segments of the facial nerve, jugular bulb, and the ICA through the cochlea, that is why complete hearing loss should be expected in the postoperative period.

A clinical case is reported of the 51-year-old female patient with trigeminal schwannoma, who presented complaining of pain in her face persisting for three months (along the branches of the right trigeminal nerve) [4]. MRI revealed a large homogenous contrasting mass in the base of the right middle fossa, extending to the most apical and medial areas of the posterior cranial fossa through the petrous apex. The patient underwent surgery via the middle cranial fossa (MCF) approach. The mass was completely resected during surgery; facial nerve paralysis with preserved lacrimal gland function, that was completely resolved six months after surgery, was observed during the postoperative period [4].

There is one more clinical case of the 33-year-old female patient with the gradually progressive facial nerve palsy (grade IV according to the House–Brackmann Facial Paralysis Scale), vertigo and nausea. Neuroimaging revealed the growing tumor that involved the geniculate ganglion and extended to the MCF, internal auditory canal (IAC) and the cerebellopontine angle (CPA) [13].

The patient undervent surgery via the MCF approach that involved the use of the facial nerve (FN) neurophysiological monitoring. The facial nerve involvement in the neoplastic process was revealed during surgery. Surgery allowed the surgeon to remove as much mass as possible and preserve the FN. The FN palsy improved gradually during the postoperative period; after 10 months of postoperative follow-up grade III of the FN function was achieved (House–Brackmann Facial Paralysis Scale), and there were no signs of tumor regrowth [13].

The middle cranial fossa (MCF) approach is used to ensure hearing preservation in patients with benign, non-tumor or cystic lesions of the petrous apex. It should be borne in mind that MCF approach to the petrous apex is used to remove, rather than just drain out the cyst; this approach is preferable when the cyst localization and abnormal hypotympanic pneumatization make it impossible to use infracochlear approach.

Middle cranial fossa (transpetrosal) approach

It proceeds from the top down towards the anterior (uppermost) surface of the pyramid and the petrous apex; the approach makes it possible to excise limited lesions. This hearing-preserving approach is unique, since it provides direct access to the petrous apex and identification of the facial nerve over a considerable length (intracranial/cisternal, meatal (mastoid), labyrinthine and tympanic segments) of the vestibulocochlear nerve and structures of the CPA [7]. Since hearing preservation is a hallmark of the MCF approach, the approach requires precision knowledge about microsurgical anatomy of the lateral skull base, given the frequent absence of the clearly defined landmarks, anatomical variability, and high risk of complications in case of surgeon's disorientation during the surgical procedure [7].

The main landmarks of the lateral skull base when using the MCF approach are as follows: middle meningeal artery, greater and lesser petrosal nerves (GSPN); trigeminal nerve (V3); projection of the IAC; arcuate eminence (EA); lacerum segment (C3 of the ICA), region of trigeminal impression.

When the lesions are large, and additional control over the carotid artery is required (when there are cysts that go down and surround the carotid artery), transcochlear access may be used, extended to the infratemporal fossa, if necessary.

Endoscopic endonasal approach

In some cases, endoscopic transnasal access can provide broad drainage pathways in patients with cholesterol granulomas of the petrous apex. The approach usually requires wide sphenoidotomy that involves creation of the large corridor to the lesion along the posterior-lateral wall of the sphenoid sinus, as well as extended maxillary antrostomy and complete ethmoidectomy in addition to removal of the posterior wall of the maxillary sinus to expose the pterygomaxillary fissure and infratemporal fossa. This approach has some advantages over transtemporal access. A significantly wider drainage tract, that is less likely to be occluded by the scar, could normally be created using transnasal approach. The disadvantages of the approach include nosebleed, liquorrhea, restenosis of the drainage tract, dry nose, complications in the form of injury to the structures of eye socket and the brain, injury to the carotid artery and facial nerve, chronic serous otitis media resulting in the need to install the tympanostomy tube; chronic sphenoiditis and transient abducens nerve palsy (cranial nerve VI), tension pneumocephalus [9,10,12].

The literature also describes the role of otoendoscopy in performing the middle cranial fossa approach. Surgical treatment of patients with cholesteatoma of the petrous apex has been assessed. The researchers have found that the use of endoscopic surgery, indicated for removal of the remaining cholesteatoma surrounding the ICA (especially in the medial part of the ICA), dura mater and facial nerve near the petrous apex, results in less invasive surgical intervention and lower number of relapses due to complete removal of the mass, especially from the so-called blind spots [14].

Meanwhile, one of the papers reports the importance of using endoscopic techniques for treatment of supralabyrinthine cholesteatoma. The techniques enabled imaging of the medial parts of the tumor surrounding the cochlea and internal carotid artery by performing transcranial approach via MCF. The 70 degree rigid endoscope was used for visualization of the internal carotid artery and the lower surface of the cochlea [3].

The case of using the combined microscopic and endoscopic approach to remove the giant petrous bone cholesteatoma is reported [15]. The authors have noted that the combined approach is mobile, it enables surgeon's manipulation in the surgical wound and provides access to the lesion from various angles.

CONCLUSION

According to literature, effusion, mucocele, cholesterol granuloma, cholesteatoma are the most common benign lesions of the petrous apex. The published sources describe various surgical approaches, among which translabyrinthine, transcochlear, endoscopic endonasal approaches and the middle cranial fossa approach are the most common.

The use of the middle cranial fossa approach, infracochlear or endoscopic transnasal approach is recommended for patients with preserved hearing. In case of disseminated lesions, when the carotid artery and the jugular bulb should be additionally controlled, transcochlear and translabyrinthine surgical corridors could be used.

Despite the wide use of endoscopic techniques in the middle ear surgery, surgery of the lateral skull base is still based on the use of microscopy, since the lateral skull base lesions are often large. It is more feasible to use lateral approaches for removal of such lesions, particularly middle cranial fossa approach, to reach the petrous apex lesion, ensure high-quality lesion removal and maximum preservation of vital vascular and neural structures, as well as the patient's hearing.

In this regard, despite all the advantages, it is better to combine endoscopic and microscopic approaches to ensure visualization of blind spots and provide additional assistance to the surgeon. The issue of the need to use endoscopic approach as a solo surgical technique remains controversial.

The middle cranial fossa approach is a hearing-preserving approach that ensures a good view of the entire petrous apex and controlled position of the facial nerve. In is necessary to have a precision knowledge of microsurgical anatomy of the bony floor of the middle cranial fossa and adjacent areas, good spatial sense, good surgical technique and sufficient experience in excisions to prevent complications when performing the middle cranial fossa approach to the petrous apex.

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