Primary nasal cavity Ewing’s sarcoma in pediatric practice: A clinical case

Relevance: Ewing’s sarcoma is an aggressive tumor rarely found in the head and neck region and extremely rare in the nasal cavity or paranasal sinuses (2-3% of all Ewing’s sarcomas).

Purpose: The article presents a clinical diagnostic case of nasal cavity Ewing’s sarcoma finally verified only by IHC test.

Results: The presented clinical case describes Ewing’s sarcoma of the nasal cavity in a 14-year-old girl and presents the clinical picture, the results of MRI and CT examination of the sinuses, histopathological and IHC tests. Adequate diagnostic tools made it possible to establish a diagnosis. Timely determined treatment tactics, taking into account the tumor localization and possible postoperative complications, led to tumor remission. At present, the patient is under dynamic observation.

Conclusion: The presented clinical case confirms that a primary nasal cavity Ewing’s sarcoma diagnostic largely depends on histopathological examination since visual diagnostic techniques do not provide reliable information on the tumor type.

Keywords: Ewing’s sarcoma, nasopharyngeal tumors, malignant neoplasms in children, pathomorphology, immunohistochemical (IHC) test.

Introduction: Ewing’s sarcoma (ES) is an aggressive tumor of childhood and adolescence. Microscopically, ES consists of small and round cells with a high nuclear-cytoplasmic index originating from primitive neuroectodermal cells. It occurs more often in early childhood or adolescence and is generally observed during the first three decades of life [1].

ES mainly arises from the bones of the skeleton: limbs, ribs, and pelvic bones. ES also occurs in soft tissues, which means fairly any possible localization in the human body. However, extra-osteal ES is rare in the head and neck area (2-3% of all ES cases) and extremely rare in the nasal cavity or paranasal sinuses [1-3].

Complaints such as anosmia, nosebleeds, snoring and hearing loss, ear pain, shortness of breath, and difficulty swallowing were described in patients with primary ES of the nasal cavity [4, 5].

Patient Information: The article presents a case of primary ES in the nasal cavity of a 14-year-old girl with complaints of breathing difficulties and right-side nasal wing edema. They consulted the ENT doctor at the Regional Advisory and Diagnosis Center; the doctor prescribed Azro, the antibiotic drug, orally. The treatment produced no response. At the next visit, the diagnosis of “Sinus cyst” was made, physiotherapy was prescribed; the physiotherapy also produced no response. Soon, the patient developed headaches irrespective to pain relievers. The patient was hospitalized. The maxillary sinus surgery, right-sided, was performed on 04.02.2020. Intraoperative examination revealed a bone defect of the anterior wall of the maxillary sinus was revealed in the form of osteolysis and a formation of 2.5x3.5 cm in the nasal cavity, easily bleeding, pale pink, dense, with a smooth surface.

A histological examination on 17.02.2020: a formation of the maxillary sinus is 2.5x3.5 cm in size, pale pink, dense consistency. Conclusion: lymphoepithelial cancer.

MRI at the residence-place clinic on 20.02.2020 showed a mass in the paranasal sinuses spreading into the oropharynx on the right and the signs of invasion into the middle cranial fossa. After the examination, the girl was consulted by an oncologist, who recommended hospitalization to the Scientific Center of Pediatrics and Children Surgery JSC, Almaty, Kazakhstan.

Clinical Data: the condition upon admission to the Scientific Center of Pediatrics and Children Surgery – severe due to the formation of the paranasal sinuses, moderate pain syndrome, signs of intoxication. The skin – pale pink, no rash. The face – moderately asymmetrical due to the swelling of the nose on the right. The mucous membrane of the oropharynx – hyperemic; an asymmetry of the soft tissues of the palate with a shift to the left due to the bulging of the peritonsillar tissue on the right. Tonsils – loose, hypertrophied.

MRI of the brain on 06.03.2020 revealed a large facial skull formation on the right, occupying the pterygopalatine and infratemporal fossa to the submandibular space, the right half of the nasal cavity nasopharynx, and ethmoid bone. The mass prolapsed into the nasopharynx, spreading into the maxillary sinus, the main sinus, and the middle cranial fossa; it did not penetrate the orbit. The tumor of irregular shape, measuring 7.0x9.8x7.5 cm, a heterogeneous cystic-solid structure, with bumpy contours, actively and unevenly accumulated the contrast solution. The nasal septum was smoothly shifted to the left by 1.2 cm; the soft palate prolapsed downward. Conclusion: Large neoplasm of the facial skull on the right (pterygopalatine and infratemporal fossa, right half of the nose, nasopharynx, ethmoid bone, and paranasal sinuses). No organic changes or metastases to the brain.

The enzyme-linked immunoassay on 03.04.2020: AFP (1.89 IU/ml) and NSE (8.4 IU/ml) – in the normal range.

Histological examination of the micro preparations on 05.03.2020 revealed neoplastic tissue of a solid structure, con-
sisting of monomorphic small cells of a round and oval shape with a high nuclear-cytoplasmic index (Figure 1). There was some accentuation of tumor cells around the vessels; the epithelial lining and glandular structures remained in some places. Focuses of necrosis and extensive hemorrhages were visualized.

An Immunohistochemical (IHC) study on 05.03.2020 showed a total positive membrane reaction with the CD99 antibody with the closure of the stained membranes (Figure 2). Reaction with Fli1 antibody revealed total nuclear staining of negative control tumor cells in stromal cells (Figure 3). The proliferative activity in the reaction with Ki67 was about 30% on a visual analog scale (not scored). Nuclear expression to INI1 in neoplastic elements was preserved; negative reaction with antibodies to PCK, EMA, EBV, CD45, CD3, CD20, desmin, myogenin, S100, CD1a, langerin, CD31, NSE, synaptophysin.
Based on the above, a tumor of the Ewing family was diagnosed according to the neoplastic tissue morphological structure and immunophenotype. A molecular genetic study to determine the translocation of the EWSR1 gene was recommended for final verification, given the rarity of this tumor localization.

A molecular genetic study on 07.03.2020 revealed the EWSR1 gene translocation. ES was finally diagnosed based on pathomorphological and molecular genetic studies, and PCT was started.

Figure 4 shows the clinical case timeline.

Discussion: Primary ES of nasal localization is mainly diagnosed by histopathological examination since visual diagnostics does not provide reliable information. MRI of tumors of this localization may suggest a malignant nature of the formation. Differential diagnostics should include malignant lymphoma, rhabdomyosarcoma, moderately differentiated carcinomas, and ES. These tumors have common X-ray signs and therefore require additional studies [6, 7].

Histologically, classical ES consists of small, homogeneous cells with scanty cytoplasm. In most cases, the nuclei are round, with depressions; small nucleoli are visualized. The cytoplasm most often has glycogen vacuoles; it can be amphophilic, with a minimum number of mitoses and stroma. ES can also include large pleomorphic cells, an organoid pattern, plump fusiform cells, and build rosettes. Changes such as nodular-alveolar structures resembling rhabdomyosarcoma, Schwann’s, and neuronal structures that mimic ganglioneuroblastomas are possible in atypical ES cases. Tissues of heterologous differentiation, such as cartilage, melanin, and muscles (ectomesenchyme), epithelial differentiation resembling adamantine are rare [8].

IHC includes reactions with monoclonal CD99, HBA71, 12E7, and FLI1 antibodies. Expression of CD99 in almost all cases is positive, with complete closure of membranes, diffusely expressed. Vimentin is always diffusely positive. Cytokeratin can be diffusely positive in the adamantine-like version [8].

Conclusions: Primary ES is very rare in the sinus tract; its diagnostics in this localization is challenging. MRI and other visual diagnostic methods are not decisive since some tumors in this localization have common radiological signs, which complicates the differential diagnosis. In this regard, a pathomorphological study of the tumor, including IHC, is the main method for ES diagnostics.

Figure 4 – Timeline of a pediatric case of primary Ewing’s sarcoma of the nasal cavity
ТУЖЫРЫМ

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Баланың мұрын қуысында өрнекласқан Юинг саркомасының алгашқы жағдайы

Өзектілігі: Юинг саркомасы – бас пен мойында сирек кездестін, ал мұрында және параназалда өзвірде сирек пайда болатын агрессивті ортаның жалпыс құрылысындағы агрессивті, злокачествен өзгілді қарайға қатысты экспандердік опухоль.

Мақсаты: Макалада мұрын қуысындағы Юинг саркомасының диагностикасы және клиникалық шарлауға болуына қарама-қарсы көрсетілді.

Өзектілігі: Баланың мұрын қуысында орналасқан Юинг саркомасының алгашқы жағдайы 14-жасындағы мұрын қуысындагы Юинг саркомасының жағдайы. Оның ортақ пайдасы – клиникалық және гистопатологиялық зерттеулер.

Заключение: Представленный клинический случай демонстрирует саркому Юинга носовой полости у 14-летней девочки, включающей клиническую картину, результаты магнитно-резонансной томографии и компьютерной томографии.

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Случай первичной саркомы Юинга носовой полости у ребенка

Актуальность: саркома Юинга - агрессивная опухоль, редко встречающаяся в области головы и шеи, и крайне редко в полости носа или придаточных пазухах носа (2-3% от всех сарком Юинга).

Цель: описание редкого случая диагностики саркомы Юинга носовой полости, ожидаемая верификация которой стала возможной только с использованием ИГХ-исследования.

Результаты: Представленный клинический случай демонстрирует саркому Юинга носовой полости у 14-летней девочки, включающую клиническую картину, результаты магнитно-резонансной томографии и компьютерной томографии.

Ключевые слова: саркома Юинга, пазуха, опухоль носоглотки, основной новый метод диагностики и лечения.