Рядък клиничен случай на хордом на околоносните кухини

A Rare Case Report of Chordoma of the Paranasal Cavities

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Резюме
Представям клиничен случай на 39-годишна пациентка (В.Т.Т.) с хордом на околоносните кухини. Извършено е радикално оперативно лечение и следоперативна лъчетерапия. В момента без данни за рецидив на заболяването. Случаят представлява клиничен интерес поради ниската честота на заболяването, трудното лечение и възможните чести рецидиви.

Ключови думи: хордома, околоносни кухини, клиничен случай, рецидив на тумора.

Summary
We present a rare case report of a 39 aged female patient (V.T.T) of chordoma of the paranasal cavities. She was treated operatively with postoperative radiation therapy up to 60 Gy. The case was interesting with very low frequency of occurrence, difficult therapy and high recurrences rate.

Key words: chordoma, paranasal cavities, case report, tumour recurrence.

Увод
Хордомите са редки тумори, изхождащи от ембрионални нотохордални остатъци. През 1857 г. Virchow пръв описва класическите отличителни патоморфоложки белези на хордома и ги е нарекъл ecchondrosis physalophora (1). Muller 1858 г. е отбелязал сходство между тези тумори и хорда дорзалис и ги е нарекъл хордоидни тумори (2). Биологичното поведение на хордомите ги характеризира като бавно растящи малигнени тумори с изразен експанзиен и деструктивен локален растеж и склонност към късно метастазиране – до 8 – 10 год. след терапията, като в повечето случаи метастазирането е след локален рецидив (3).

Introduction
Chordomas are rare tumours originating from embryonal notochordal remnants. In 1857 Virchow was the first to describe the classic distinctive pathomorphological characteristics of chordoma. He called them ecchondrosis physalophora (1). In 1858 Muller noted the similarity between these tumours and the notochord (chorda dorsalis) and called them chordoid tumours (2). Chordomas’ biological behaviour characterizes them as slow-growing malignant tumours with expressed expansive and destructive local growth and tendency towards late metastatic processes – up to 8-10 years after the therapy- the metastases usually developing after a local tumour recurrence (3).
**Epidemiology**

Chordoma’s frequency of occurrence is 1 to 8÷10 000 000. It represents 1% of the primary bone tumours and approximately 3% of the primary malignant bone tumours. Gender distribution: men are affected twice as often as women. Chordomas occur predominantly in adults; most frequently in patients aged forty to sixty. They are very rare in young people, the percentage being < 5% in patients under the age of twenty.

**Localization**

Chordomas occur mainly along the length of the neuroaxis and are most frequently observed in sacrococcygeal region (50 – 60%), spheno-occipital region (35%), shiynata zona (10%) and torak-o-llumbal region (5%) (5, 6). Although the huge percentage of chordomas affects the axial skeleton, there have been reported rare cases of extra-axial chordomas. Ectopic chordomas genesis presupposes dispersed chordarests, appearing in the whole region of the scull. There have been observed patients whose nasal and paranasal cavities and orbits have been affected.

**Clinical Case**

V.T.T, aged 39, female (Ref. 7617/27.08.2014). The patient was referred to the Department of Ear, Nose and Throat Diseases by an ophthalmologist. The patient complained of a right eye angle swelling and epiphora having lasted for a number of years. The diagnosis on hospitalization was „Dacryocystitis vдясно“. Objetively se установява туморна формация в областта на слъзната кост с твърдо-еластична консистенция, неболезнена и без промяна в надлежащата кухина. Размери: 20 х 10 мм. Планува-на интервенция: DCR dex. The computed tomography scan of the nasal cavities revealed an expansive soft tissue formation in the right aspect of the ethmoid complex, destroying the medial orbit wall and invading part of it. CT scan exam diagnosis: mucocele (Fig. 1).

**Surgical Treatment I**

On 28.08.2014 an endoscopic endonasal sinus surgery was performed under general anesthesia. A tumour formation, engaging the right nasolacrimal duct and having maxillo-ethmoidal nodular macroscopic structure and hard-elastic consistency was removed. Histological test No 4061, 4062/14B – chordoma.
Post-operative Study

Three months later (Ref. 10493/04.11.2014) clinical and CT scan findings indicated tumour persistence. A formation with heterogenic structure and 20-37 XE densinometric characteristics was visualized in the upper medial edge of right maxillary sinus. The bone structures in the region were destructed. CT scan conclusion: Tumour formation (Fig. 2).

Figure 1. In the right aspect of the ethmoidal complex an expansive soft tissue formation is observed, which is seen to destruct the medial orbit wall and invade part of it. CT scan exam diagnosis: mucocele.

Figure 2. A formation with heterogenic structure and 20-37 XE densinometric characteristics was visualized in the upper medial edge of right maxillary sinus. The bone structures in the region were destructed. CT scan conclusion: Tumour formation.

Surgical Treatment II

On 05.11.2014 excision of the tumour formation in the inner eye corner on the right and the anterior medial wall of the right maxillary sinus was
performed through a combined endoscopic endonasal access and an additional vestibular incision in the gingiva-buccal sulcus. Histology: chordoma fragments. The Oncological Committee decided on post-operative radiation therapy. During the radiation therapy conducted a persistent tumour formation was found again. CT Scan findings: the right maxillary sinus has destructive changes along the medial wall and is totally engaged by a mixed density formation of 17 to 25 XE. Partial engagement of the ethmoid complex is visualized. Bone destruction is observed in the medial edge of the right orbit as well.

**Surgical Treatment III**

On 18.03.2015 (Ref. 3386/17.03.2015) under general anesthesia, with Moure’s incision and a lateral rhinotomy on the right, a medial maxillectomy with excision of the tumour and resection of the anterior orbit and medial wall of the right maxillary sinus with a part of the ethmoid labyrinth was conducted. Histology: chordoma. Decision was taken for radiation therapy up to 60 Gy. Postoperative follow-up show changes in the region of the right maxilla-ethmoidal angle and presence of polypoid growth without tumour recurrence (Fig. 3).

**Figure 3.** CT scan – post-operative changes in the region of the right maxilla-ethmoidal angle and presence of polypoid growth.
Discussion

Chordomas are neoplasms originating from embryonal notochord and are generally localized medially in the region of the craniovertebral axis. Under macroscopic study the tumour has a nodular appearance /Tsenev, 1997/ with uneven surface, whitish-yellow colour and hard-elastic consistency with areas of haemorrhage (8). There are 3 histological variations: classic/conventional; chondroid chordoma; dedifferentiated variation. The macroscopic study showed that the tumour is similar to the chordal tissue with the accumulation of large physalis shaped (balloon-like) cells with a small, pushed to the periphery nucleus. The intercellular space is filled with protein-musinosis substances. The modern diagnosis is based on immunohistochemistry and includes a positive reaction for epithelial markers: Cytokeratin (+++), Epithelial membrane antigen (EMA) (+++), S-100 protein (+++) and Carcinoembryonic antigen (+) (9).

Factors predetermining the unfavorable prognosis are: tumour volume, old age, female gender, some histological and cytological markers: presence of tumour necroses in a volume above 10%, abnormal mitotic index, chondroid differentiation of the tumour (Forsith et al. 1993) (2).

Chordomas require complex treatment including mainly surgical removal and different types of (pre-operative, post-operative, definitive) radiation therapy.

Even the most radical intervention cannot guarantee complete healing. Recurrences are always possible in a long-term patent’s monitoring. The results from chordoma’s surgical treatment vary widely and depend on different factors:
1. Extent of tumour resection – maximum radical intervention is the main factor for remission duration;
2. Local status – tumour size and location, resurgery, preceding radiation therapy.

Post-operative adjuvant radiation therapy is used in an attempt to be achieved local tumour control, prevention and slowing of recurrence development, and, if possible, better survival (10). The results of radiation therapy depend on the volume of preceding tumour resection. Due to the relative resistance of chordomas to radiation therapy, the extent of
resistant to radiotherapy. The volume of tumour excision is the main factor determining the treatment outcome (2).

Pre-operative radiation therapy aims at decreasing the tumour volume and makes its removal possible. Definitive radiation therapy is conducted to inoperable patients (3). There are no convincing data for a significant effect achieved as a result of chemotherapy conducted as chordomas are resistant to the cytostatic drugs (11).

5-year patients’ survival rates vary around 60-70%; ten-year studies reveal patients’ survival rate of 30-40% (Dorfman 1998). Donald et al. report average survival rate of 51% for a five-year period. Local-regional recidivisms are considered to be the leading cause for a fatal outcome (5). Despite the possibility of distant metastases, local recurrences are the most frequent reason for unfavourable prognosis. Distant metastases (lungs, bones, soft tissues, lymph nodes, liver and skin) vary from 0 – 40% (Chambers et al. 1979) to 40-60% (Cotton 1996) (3, 5).

Development of chordoma metastases in most cases occur late in the disease evolution, usually after a local recurrence. The prognosis is related rather to the local tumour aggressiveness than to the metastatic process.

Study

Regular out-patient observation aims at early detection of potentially treatable local-regional recurrences and the presence of disease dissemination. The study of patients includes medical examinations, use of imaging and endoscopic techniques. It is recommended that patients’ study be conducted every three months during the first 2 years. Patients should be examined every 4 months during the fourth year, and every 6 months in the period 5-10 year (3, 12). All operable recidivisms are subject to most radical surgical removal.

Conclusions

1. Chordomas are rare primary tumours with a high risk of local recurrence and late predisposition to distant metastases.
2. Surgical intervention is a method to be considered when choosing a treatment method.

Продължаване

Регулярното диспансерно наблюдение цели ранно откриване на потенциално лечим локорегионален рецидив, както и наличие на десиминация на заболяването. Проследяването на пациентите включва преглед, обрани и ендоскопски методи. Препоръчва се първите 2 години проследяването да се извършва на всеки 3 месеца, през 3-та и 4-та година на 4 месеца, а от 5 до 10 години на всеки 6 месеца (3, 12). Всички операбилни рецидиви подлежат на максимално радикално оперативно отстраняване.

ИЗВОДИ

1. Хордомите са редки първични костни тумори с висок риск за локален рецидив и възможно предрасположение за далечни метастази.
2. Хирургичната интервенция е метод за избор за лечение на заболяването.
3. Post-operative radiation therapy plays a significant role for local control of disease.
4. Active and result-oriented study allows early detection of recurrences and metastases; their treatment improves the prognosis.

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