Intracranial Dysontogenetic Tumors in Children: Topographic-Histologic Correlations, Own Experience and Review of the Literature

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Abstract

Introduction: Dysontogenetic tumors (DTs) are developmental disorders, resulting from displaced secreting skin epithelium, manifesting by cysts filled with cellular detritus, cholesterol, sebum, hair, etc. DTs generally include epidermoid cysts (ECs) and dermoid cysts (DCs), but also colloid cysts, enterogenous cysts, mature teratomas and cranioopharyngiomas. This paper will focus on ECs and DCs only.

Aim of Paper: The aim is to present own experience in the treatment of intracranial DTs in children, clinical-topographic-histologic correlations and problems encountered.

Materials and Methods: The Study group included 21 children (14 males, 7 females) aged 6 months - 18 years (mean age 9.1 y) with a histologically verified DT, treated at our institution since 2004 thru 2014. Mean follow-up was 5.7 y. Retrospective analysis of medical records was the technique used.

Results: The Study group included 12 ECs (8 m, 3 f) and 9 DCs (6 m, 3 f). The factors analyzed were gender, age at first diagnosis, clinical symptoms, location of lesion, histological profile and presence of a dermal sinus. All patients underwent surgery without perioperative mortality. Late outcome was favorable in 15/21 cases (71%) and poor in 6/21 (29%).

Conclusions: Intracranial DTs are more frequent and take a more aggressive clinical course in males, with a predominating anterior location. DCs more often arise near the midline and coexist with a dermal sinus. All patients underwent surgery without perioperative mortality. Treatment outcomes are mostly favorable but peri- and postoperative complications may result in permanent functional and cosmetic sequels.

Keywords: Dysontogenetic Tumors; Epidermoid Cysts; Dermoid Cysts; Intracranial

Introduction

Dysontogenetic tumors (DTs) are a highly heterogenous group of pathologies in terms of clinical signs and course, histopathology and prognosis. Depending on location and organs involved, they pose a challenge for neurosurgeons, dermatologists and cosmetologists, general surgeons, gynecologists, pediatricians, ophthalmologists, ENT specialists and, ultimately, pathologists, if only due to ambiguous diagnostic criteria and inconsistent nomenclature. DTs of virtually any location have been described, including orbit [1], intracranial [2], intralingual [3], parotid [4,5], floor of the mouth [6], testes [7], ovaries [8], omentum [9] and nose [10] to name just a few.

Essentially, these are congenital defects, resulting from the presence of misplaced secreting skin epithelium, forming cysts filled with residues of desquamated cells and products of secretion of epithelial glands eventually present therein. On traditional grounds, the term “DT” encompasses epidermoid cyst and dermoid cyst, but in some authors’ opinion this group should include also colloid cysts, neuromeric cysts, Wilkins and Odum’s grade I teratomas and even cranioopharyngiomas. For the sake of clarity of presentation, in this paper we will address the first two categories only.

The Pathogenetic significance of DTs results from mass effect, highly irritating cyst content (cholesterol crystals, semi-liquid lipids and keratin detritus), relatively high recurrence rate if incompletely excised and high risk of CNS infections due to a frequent communication between the lesion itself and skin surface. Furthermore, lesions located in exposed and cosmetically important areas, e.g. face, scalp or lumbo-sacral area, may be a source of much discomfort and distress for patients and their relatives.

DTs pose a considerable clinical challenge. Being a congenital pathology, they are usually intimately bound with adjacent structures, sharing vascular supply and lacking a clear anatomical dissection plane. Attempts at complete surgical excision may result in a damage of adjacent structures, leading to devastating loss of function. Surgical manipulation of a DT harboring latent infection may result in a severe and treatment-resistant local or generalized infection. On the other hand, non-radical excision may result in a recurrence with every subsequent procedure being more traumatic and less effective.

An additional problem is the mere rarity of DTs, precluding accumulation of clinical data enabling development of optimal management standards.

Aim of Paper

Our aim was to present our experience in the management of intracranial DTs in children, highlighting regularities observed and problems encountered. As mentioned above, in order to reduce heterogeneity of the group, we limited our analysis to DTs in the traditional terms, i.e. epidermoid cysts (ECs) and dermoid cysts (DCs).

Materials and Methods

Study population consisted of 21 children diagnosed with histologically confirmed DTs, treated at the Department of Neurosurgery, Children’s Memorial Health Institute, Warsaw, Poland, over ten years’ period - since 2004 through 2014. The group included 14 boys and 7 girls aged from 6 months to 18 years (mean age: 9.1 y). Mean follow-up time was 5.7 y. The study was performed by retrospective analysis of medical documentation.

Results

Our material (DTs, n = 21) included 9 DCs (6 boys and 3 girls)
and 12 ECs (8 boys and 3 girls). Noteworthy is the prevalence of males in both tumor types. Furthermore, the mean age at presentation was 8.8 y in the males vs. 12.5 y in the females, indicating a more aggressive course of DTs in the former, resulting in earlier development of clinical signs.

The entire study population was subdivided depending on location of the lesion, which is the key factor determining clinical signs, treatment strategy and outcome, i.e. anterior fossa, orbit, lateral sulcus, III-rd ventricle and pineal area, cerebellum and cerebello-pontine angle.

“Anterior fossa” group included 5 boys and 1 girl, harboring 3 ECs and 3 DCs, thereof 5 presented with a dermal sinus. Brain abscess was the initial diagnosis in one case and septo-optic dysplasia in one case. Mean age at presentation was 6 y.

“Orbital” group included 3 boys, harboring ECs only. The leading sign was exophthalmos (2 cases) and strabismus with diplopia (1 case). Mean age at presentation was 8.5 y.

“Lateral sulcus” group included 2 boys and 2 girls, harboring 3 DCs and 1 EC. The leading symptom was epilepsy (3 cases) and headache (1 case). Mean age at presentation was 11.2 y.

“Cerebello-pontine angle (CPA) group included 3 boys and 2 girls, harboring ECs only. The Presenting symptoms included cranial nerve palsies (IV, V, VII, VIII and IX) and epilepsy (1 case). Mean age at presentation was 15.6 y.

“Posterior fossa” group (excluding CPA) included 1 boy and 1 girl, harboring DCs only. Presenting signs included dermal sinus (both cases), subcutaneous abscess, bulbar syndrome and hydrocephalus (1 case each). Mean age at presentation was 3 y.

“Pineal area and third ventricle” location was seen in 1 girl with an EC, manifesting by headache. Her age at presentation was 15 y.

Overall, dermal sinus was present in 2/12 of ECs (16%) and in 5/9 DCs (55%). Considering correlation between location of lesion and dermal sinus, it was present in 5/6 (83%) of anterior fossa lesions (3 DCs and 2 ECs) and in 2/2 (100%) of posterior fossa lesions (DCs only). Based on our material, it is clear that dermal sinus is mostly associated with midline lesions and with DCs.

All patients underwent surgery, thereof 20 achieved radical resection and 1 – non radical (a boy with a nasal dermal sinus, presenting with an abscess of the left frontal lobe and a DC adjacent to the cribriform plate and penetrating his left frontal lobe).

Postoperative course was uneventful in 17 cases (80%) and complicated in 4 cases (20%). Encountered problems included SIADH (EC in the hypothalamic area), malignant edema of subependymal structures (EC in the pineal area), recurrent abscess of brain and soft tissue (DC of the frontal lobe) and meningitis (DC of the posterior fossa). Noteworthy is that EC-related complications resulted mostly from location of the lesion and damage to adjacent brain, while DC-related problems were mostly infectious in nature.

Long-term outcome was favorable in 15 cases (71%) – patients are currently neurologically intact without any disease-related ailments. In the remaining 6 cases (29%), the outcome is unsatisfactory. Permanent sequels of the disease and its treatment

Figure 1: Location of intracranial DTs present in females. (Ovals – ECs; triangles – DCs; arrows – associated dermal sinus).

Figure 2: Location of intracranial DTs present in males. (Ovals – ECs; triangles – DCs; arrows – associated dermal sinus).

Figure 3: Location of DTs located within the spinal canal. (Ovals – ECs; triangles – DCs; Arrows – associated dermal sinus).
Intracranial DTs are essentially a congenital defect, caused by ectopic location of embryonal ectoderm inside or adjacent to an already formed neural tube during 3–6th gestational week. EC is lined by keratinized stratified squamous epithelium and filled with desquamated epithelial cells, keratin and cholesterol, forming a whitish, brittle mass on gross inspection. It does not contain dermal appendages (e.g. hair or sebaceous glands). DC is also lined by multilayered squamous epithelium, but it does contain skin adnexae: hair follicles, sweat and sebaceous glands, and enamel buds. It is filled with a yellowish, oily mass, containing hair and sebum. Rare cases of acquired DTs have been described, developing as a consequence of a beesting, liposuction, lumbar puncture and other situations resulting in an accidental displacement of dernal tissue.

An important DT-related issue is the highly irritant effect of cyst content repeatedly released into the subarachnoid space. According to the literature, about 7% of DTs rupture spontaneously, leading to aseptic meningitis and vasospasm, which may be fatal or may give rise to acute hydrocephalus with potentially lethal elevation of intracranial pressure.

DTs may give rise to various skin-related neoplasms (melanoma, squamous cell carcinoma, basal cell carcinoma, trabecular carcinoma) further complicating the situation. Several reports highlight the role of human papillomavirus in the development of DT-related tumors and presence of carcinoembryonic antigen in affected persons. Cancerogenesis may be heralded by rapid enlargement of a hitherto stable lesion leading to aseptic meningitis and vasospasm, which may be fatal. Potential malignant transformation is additional indication for surgical treatment of DTs. Intense pigmentation of these lesions has also been described.

There are no blood vessels inside a DT cyst. If cyst content becomes infected by a co-existing dermal sinus, it acts as an infected alien body. In such a setting, conservative control of the infection is extremely difficult if not impossible.

Congenital DTs are intimately bound with adjacent normal structures, often lacking a clear plane of cleavage and sharing vascular supply. Recurrent discharge of irritant cyst content enhances chronic inflammation and scarring, further complicating surgical excision. The cornerstone of successful and complication-free therapy is precise topographic diagnosis, careful preoperative planning and complete excision (if feasible). Perioperative antibiotic prophylaxis is mandatory. An important issue is proper timing of surgery, i.e. avoiding operation during active infection of lesion or periods of immunologic compromise of the host.

Recurring DTs located within the spinal canal may be treated by radiation therapy. Several reports confirm viability of this option. In our material, we have one such child (boy, aged 8 with a DC at L3-S2 level. One year after radiotherapy his lesion appears stable and neurological condition is satisfactory (no ailments, ambulant, normal vital activity).

To sum up, while essentially benign and rare lesions, DTs deserve attention due to the complexity of anatomical, functional and cosmetic issues associated therewith, as well as potentially devastating consequences if left untreated. Furthermore, difficulties encountered during and after surgery are arguments for referral of such (fortunately) rare cases to highly experienced and skilled centers for treatment.

**Conclusions**

1. Intracranial DTs are more frequent and progress more rapidly in the males, resulting in earlier manifestation. Predominates in anterior location of the lesion (anterior fossa and orbit).
2. DCs develop mostly at or near the midline and are more often associated with a dermal sinus.
3. ECs develop more laterally and rarely coexist with a dermal sinus.
4. Symptoms depend on location and coexisting dermal sinus, promoting intra- or extracranial infection.
5. Treatment outcomes are usually favorable, although a complicated postoperative course is to be expected.

**Conflict of Interests**

There is no conflict of interest among authors.

**References**