GIGANT FACIAL NEUROFIBROMA

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In August 2003, a 15 year old male, with a giant tumor located on the whole left side of his face, was admitted for treatment. The changes in appearance first began to show in the 2nd month after birth and systematic enlargement lead to an overgrowth of the soft tissues and bones of the face. As a result of the pressure exerted by the tumor, the frontal bone and the surrounding protective structures became thickened and displaced posterio-inferiorly, causing a shrinking of the anterio-inferior part of the skull. Histological analysis indicated that the tumor was of neurological origin. The patient was operated on a year later because of the continued growth of the tumor. Another year later, the enlarged and protruding maxillar bone was filed down.

A radical approach is important in such cases, not only from an aesthetic point of view, but most of all because it is the best way of counteracting any regrowth of the tumour and is the most effective means of oncological prophylaxis.

Key words: neurofibroma, Recklinghausen disease, face tumour

Neurogenic tumours of the head and neck constitute about 2% of all tumours occurring in this region (1). They usually arise from peripheral nerves, ganglia, paraganglionic cells and embryonal ectoderm.

According to Laudreneu’s classification (2), neurofibromas belong to tumours arising from peripheral nerves. They most commonly occur in the form of type 1 neurofibromatosis (NF-1), referred to as von Recklinghausen disease, or type 2 neurofibromatosis (MISME Syndrome- Multiple Inherited Schwannomas, Meningiomas and Ependymomas). Both types are genetically conditioned and originate as a result of chromosomal mutations – chromosome 17 in case of type 1 and chromosome 22 in case of type 2 neurofibromatosis.

In Recklinghausen disease tumours of various sizes are disseminated on the skin all over the body. On the face the patches may combine and form larger conglomerations (3). They are often accompanied by light brown patches in the form of café-au-lait spots or freckles (4, 5).

The most characteristic feature of NF-2 are bilateral neurofibromas on nerve VIII observed in 90% of cases. Almost half of the patients also develop meningioma and changes within the spinal cord.

Isolated facial tumours without other typical symptoms of neurofibromatosis are rare (6, 7). Cases of elephantiasis neurofibromatosa, in which facial tumours assumed huge size due to progressive hypertrophy of fibrous and nervous elements of the peripheral nerves have also been described (8, 9, 10).

CASE REPORT

Patient M.L., age 15, was admitted to hospital in August 2003 for treatment of a giant tumour covering the whole of the left face. The lesion appeared in the 2nd month of age and
increased systematically, leading to hypertrophy of the soft tissue and facial bones. Throughout the whole period the patient did not receive any treatment, however he was attended at the Oncological, Neurological and Maxillofacial Out-patient Departments in one of the academic centres.

On admission he presented with a huge tumour involving almost whole of his left face. The tumour involved the whole temporal fossa from above, extended anteriorly to the forehead, posteriorly to the parietal bone and superiorly beyond the border of the temporal bone squama. The central part of the tumour was localized to the region of the zygomatic bone and covered the maxillary prominence, it reached the midline on the nose and extended posteriorly toward the auricle margin. The lower border of the tumour extended toward the inferior margin of the mandible, covering the mandibular ramus, angle and major part of the body on the left side. Huge size and weight of the tumour caused distention of the external angle of the left eye and deformity of the whole palpebral fissure as well as downward dislocation of the mouth and deformity of the nose (fig. 1).

The tumour was uniformly solid all over its surface, slightly yielding to pressure and slightly movable. The underlying skin was tense, and in the region of ala nasi it revealed trophic lesion covering the surface of 2 x 5 cm.

CT scan revealed extensive changes within the cranial bone. Due to the pressure exerted by the tumour mass, the frontal bone and temporal squama were thickened and displaced posteriorly and medially. The size of the anterior cranial fossa on the left side was reduced in all the dimensions, especially the width. Bones forming the lateral orbital margin were narrowed and the lateral wall was thinner and flattened. The zygomatic bone body was thickened and the temporal process of the zygomatic bone was hypertrophic and displaced downwards. The whole maxillary bone was hypertrophic, especially in the region of the body and the frontal process with narrowing of the maxillary sinus and paranasal sinuses on the affected side.

The general condition of the patient was good; apart from the above-described deformities he did not present any other deviations from the norm. Limitation of the visual field, problems with ingestion of food and difficulties in speaking were the consequence of direct pressure of the tumour mass on neighbouring tissues.

The operation was performed under general anaesthesia. The skin overlying the tumour was injected with 0.5% solution of lignocain with addition of adrenalin. The skin overlying the whole tumour was extensively dissected from incision running from the mandibular angle upwards in front of the auricle on the temple and along the hairline on the forehead. Moving backwards, the tumour was separated from the background. Operation on the skull was relatively easy due to a distinct border between the tumour and the periosteum and the bone. Lack of the capsule and overgrowth of the neurofibroma structures into the underlying facial muscles hampered the course of the procedure due to profuse bleeding. The tumour was resected almost completely, only small part of neurofibroma mass located medially below the orbit in the region of ala nasi remained (11). In this place the border between the tumour and healthy tissues was especially blurred.

Histopathological examination of the resected tumour revealed neurofibromatosis. Histochemical examinations S-100 and NSE pointed to neurogenic origin of the tumour. Low proliferation index Ki-67 indicated benign character of the tumour.

The patient was readmitted after 6 months due to slowly progressing, steady growth of neurofibroma mass, mainly on the cheek and in the pre-auricular region. Using previous approach, the tumour was extensively resected until the healthy tissues margin on the nose and in the region of medial angle of the eye. The hypertrophic and augmented maxilla was filed with a cutter. In order to correct the hollow in the temple, it was filled with rescored bone fragments and corium graft obtained by de-epithalization of the removed excess skin covering the tumour.

Histopathological evaluation of numerous specimens revealed only the presence of neurofibroma or neurofibroma pigmentosa.

The patient was readmitted again in October 2004 for hyperthrophy of the tumour, which projected to the oral vestibule at the level of molar teeth. The pathological tissue covering the maxilla at the width of 6-7 cm and reaching the infraorbital foramen was excised from the intraoral approach.

The histopathological examination confirmed previous diagnosis.
Fig. 1. Giant facial neurofibroma.
A, B, C – giant tumour in a 14-year-old patient; D, E – computer pictures of the facial skeleton; F – neurofibroma projecting to the oral cavity and deforming the maxilla; G – facial flap dissected over the tumour – intraoperative picture; H – excised neurofibroma size 22 x 18 x 7 cm; I, J, K – outcome of the surgery
The patient was operated on for the last time in June 2005. The surgery consisted in removal of tissue in the region of lateral wall of the nose from the approach through the former scar in the oral vestibule. The whole thickened and augmented surface of the frontal process and maxilla body was filed until the level of inner table of the bone.

RESULTS

Almost complete excision of the giant tumour extending over the whole left side of the face changed radically the patient’s appearance and the previously deformed face assumed almost normal shape. The only remain was a slight asymmetry of the left orbital margin, projection of the maxilla and dislocation of the zygomatic arch, which resulted either from bone hypertrophy or from the pressure of the tumour mass on the neighbouring bone, which had lasted for 15 years. Another consequence of all the operations was nerve VII paresis – slight dropping of the mouth angle associated with paralysis of the frontal branch. Extensive scar in temporal and preauricular area was not a problem, as the patient managed to conceal it effectively.

DISCUSSION

Luckily, giant facial neurofibromas occur rarely. For this reason the management has to be determined individually in every case depending on the size and localization of the tumour as well as on the skills and possibilities of the operating team. The radical character of the procedure is associated not only with the cosmetic aspect. As was shown in our case, even small mass of tumour (less than 10% of the initial mass) left in place resulted in rapid proliferation. Therefore, maximally radical excision is the best method of oncological prophylaxis, although in such cases complete removal of the tumour is rather impossible (12-15).

In our patient we managed to avoid extensive damaging of the facial nerve, however it may occur to be an unavoidable complication of similar procedures in other cases. We hope that the patient’s occlusion and facial asymmetry can be ameliorated with recently undertaken orthodontic treatment and planned refinement procedures.

CONCLUSIONS

1. The therapy of isolated facial neurofibromas should be initiated early, even at the pre-school age, in order to prevent giant hypertrophy.
2. Tumour resection should be as radical as possible, depending on the anatomical conditions.

REFERENCES


Received: 18.06.2008 r.
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