Nasal glioma

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The nasal glioma was first described by Reid in 1852. Its genesis was thoroughly discussed by Schmidt (1900) and up to 1966 ninety-six cases had been reported, including those reported on in more extensive surveys (Black and Smith, 1950; Walker and Resler, 1963; Smith *et al.*, 1963). The most usual term for this growth is nasal glioma, but a number of synonyms are encountered: encephalochoristoma nasofrontalis (Zettergren, 1948), intra-nasal meningocele (Hallerman, 1932), congenital ganglioneuroma (Crook, 1940).

The majority of these nasal gliomas appear at, or shortly after, birth (Black and Smith, 1950; Walker and Resler, 1963; Smith *et al.*, 1963). They are generally situated extra-nasally (60 per cent), but they also occur intra-nasally in 30 per cent of cases and both intra- and extra-nasally in about 10 per cent (Matheson, 1964). When they appear later, the order is reversed, as can be seen from the surveys by Black and Smith (1950) as well as Smith *et al.* (1963). Wang and Macomber (1964) state that the male : female sex ratio is 3 : 2. No familial predisposition has been noted; nor has a higher incidence of other anomalies been observed (Walker and Resler, 1963).

The extra-nasal gliomas generally appear as solid, usually subcutaneous, tumours astride, or on either side of, the mid-line through the root of the nose, while the intra-nasal ones are encountered laterally in either nasal cavity, sometimes associated with dislocation of the septum and nasal bone, and covered by respiratory tract mucosa (Black and Smith, 1950; Ash *et al.*, 1964; Evans, 1966). Macroscopic descriptions are rare, but the nasal glioma is described in some cases as being grey, yellowish or purple in colour and of a fairly solid consistency (Smith *et al.*, 1963; Walker and Resler, 1963; Christianson, 1966). It is seldom recognized macroscopically as cerebral tissue. Histologically, nasal gliomas consist primarily of astrocytes, which are not infrequently large and characterized by irregular, and sometimes multiple, nuclei. Mitoses are extremely rare and genuine neuronal cells are usually not present, although they have been reported. Various degrees of fibrosis and the existence of blood vessels are seen (Zettergren, 1948; Ross, 1966). An electron-microscopic investigation has been described in one case (Smith *et al.*, 1963). The rate of growth is the same or slightly increased compared with the normal nasal tissues. The most rapid rate of growth in Black and Smith's survey (1950) is 'obstruction of nose in 7 weeks'. Recurrences after extirpation are reported in five patients in their summary of 36 cases. The earliest recurrences appeared after one month and the latest after 35 months. A number of theories have been advanced concerning the pathogenesis of the tumour. Most of these are carefully noted, with an indication of the sources, in the three more extensive surveys by Black and Smith (1950), Walker and Resler (1963) and Smith *et al.* (1963). Consequently, a detailed account will not be given here. However, according to Smith *et al.*

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(1963) it is generally accepted that nasal gliomas have the same type of origin as encephaloceles. From the third to fifth embryonal week the posterior neuropore is open and is connected with the nasal cavity by a cord of epithelial cells. Herniation of the brain can occur along this opening. The meninges and cranium can then close behind the herniation, leaving an isolated mass of neuroectoderm. If the closure is incomplete, a defect develops in the cribriform plate and a connection between the nasal glioma and the intracranial structures persists in the form of a connective tissue cord that sometimes also contains glial tissue. These defects in the base of the skull are extremely difficult to diagnose other than by anterior craniotomy, and even when a defect has been demonstrated by this method, it has not been possible to detect it on subsequent examination of a straight X-ray or laminagram. Pneumoencephalograms have seldom been used, and positive findings have been reported only in two patients (Smith *et al.*, 1963). Ackerman (1968) regards the nasal glioma as a bridge between purely anatomical defects on the one hand and low-grade neoplasia on the other. In some of the cases published hitherto, recurrences have been noted and the glioma has grown more rapidly than other tissues. Invasive growth or the formation of metastases has not been reported. From the point of view of differential diagnosis, Wang and Macomber (1964) have set up three main groups to which tumours and tumour-like growths of the nasal cavity can be assigned: those of neurogenic origin (gliomas, encephaloceles, neurofibromas), those of ectodermal origin (dermoid cysts, nasal polypi and cysts from sinuses and lacrimal ducts) and those of mesodermal origin (haemangiomas, lipomas).

The differential diagnosis of the nasal glioma as opposed to a genuine encephalocele or meningocele may prove difficult. In the latter case there are connections with the brain cavities containing cerebrospinal fluid, and the risk of post-operative meningo-encephalitis following extirpation increases very considerably. The leakage of cerebrospinal fluid during surgery is an important sign and should be followed up with craniotomy (Smith *et al.*, 1963; Ross, 1966); Walker and Resler (1963) suggest that craniotomy should always precede other operative measures against nasal glioma.

Case report

The patient is now a 4-year-old boy, the second child of a healthy bipara, born by normal delivery after an uncomplicated pregnancy with a birth weight of 3,760 g. No deformities were noted in the patient, nor were any known to exist in his family. When he was one month old, a marked right-sided nasal obstruction was noticed and the parents sought consultation at the ear clinic at Södersjukhuset (Southern Municipal Hospital) in Stockholm. On inspection a lateral displacement of the nasal bone to the right was observed, as well as a rather soft, somewhat bluish mass that filled the right nasal cavity. It was attached by a broad base on the lateral wall and covered by normal mucosa. At fine-needle aspiration biopsy, only a clear, mucous fluid and cellular elements from the nasal mucosa were found and the mass was judged to be cystic.

A decision was made to try to extirpate the pathological tissue and the patient was operated upon at the age of three months. The ala nasi was freed and folded upward. The inferior portion of the mass was readily disengaged,

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but towards the ethmoidal area the extirpation probably was not total. During the course of the operation we got the impression that the pathologically changed tissue was a homogeneous tumour and not a cyst. Good mucosal coverage was easy to achieve. No leakage of cerebrospinal fluid was noted. The patho-anatomical diagnosis revealed highly differentiated brain-like glial tissue with respiratory tract epithelium and isolated glands: a nasal glioma (Fig. 2). The day after the operation the patient had a rise in temperature; lumbar puncture revealed sterile cerebrospinal fluid with normal cell and protein contents, and the patient quickly became afebrile. Subsequently, the child was transferred to the Sachska Childrens' Hospital for further investigation. X-ray of the skull and facial bones were taken and showed normal conditions. On pneumoencephalography no filling of the cisterna lamina terminalis and the posterior portion of the subfrontal region was noted, and therefore, right-sided angiography was performed. The examination showed normal conditions. The funduses oculi were normal. A certain degree of non-specific

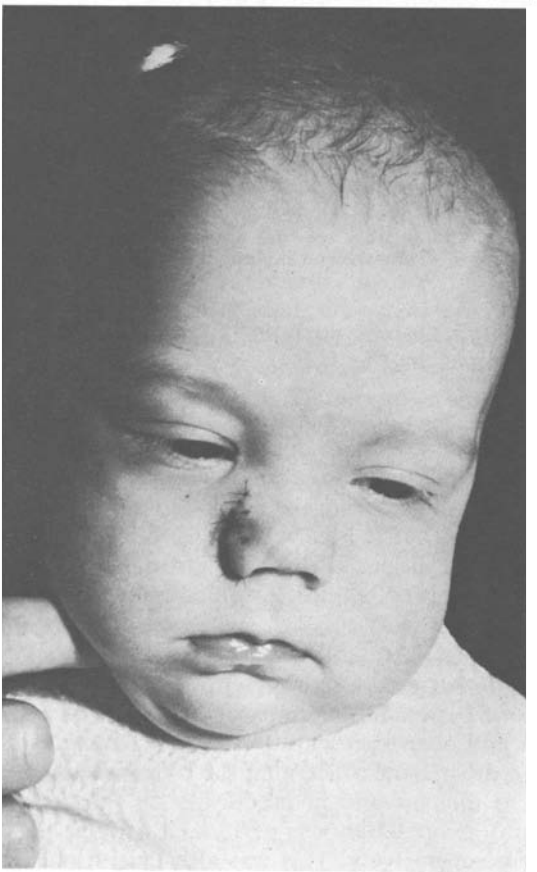


FIG. 1.
Picture showing the case described four days after operation. The configuration of the nose is the same as before the removal of the tumour.

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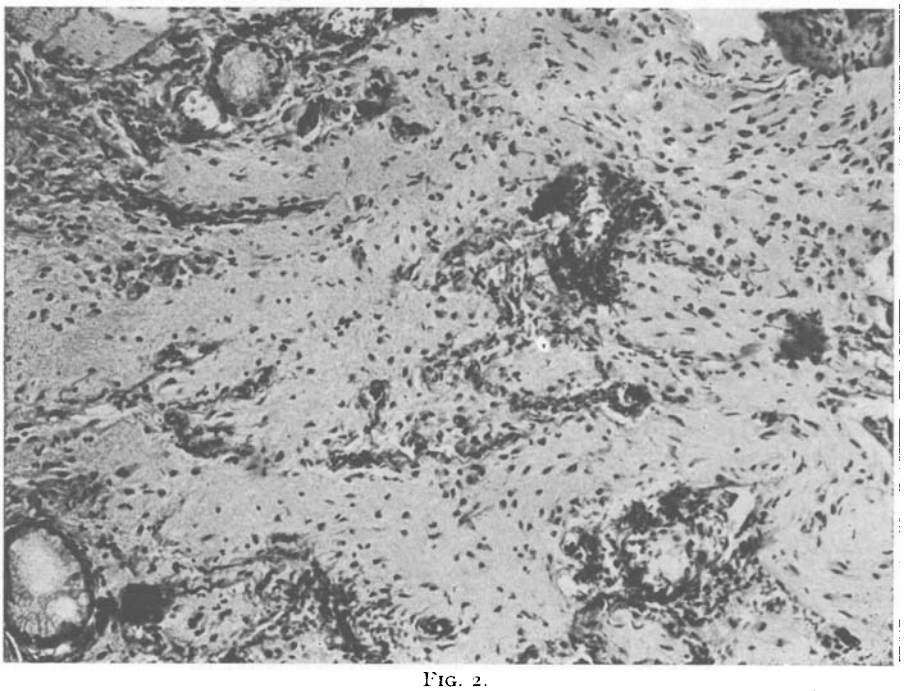


FIG. 2.
Histologic view of nasal glioma from the case described showing astrocytic glia, fibrosis, capillary vessels and one mucous gland incorporated. van Gieson, green filter. ×125

asymmetrical activity, most pronounced on the left side, was recorded from the post-central regions in the E.E.G. X-rays of the nasal sinuses showed normal conditions.

The child has now been observed for a period of four years and is entirely free from discomfort and without neurological symptoms. Nasal patency is very good and the mucous membrane is normal. A slight lateral thickening was detected in the nasal cavity during the first two years, but this has diminished during the last year so that it is now only barely discernible. There is still a certain degree of lateral dislocation of the right nasal bone, but it is so slight that it can only be observed on close examination.

Discussion

Nasal gliomas are extremely rare growths, which, owing to their nature of an intermediary form between a deformity and a tumour as well as their location, neurogenic origin and over-representation in very young growing individuals, always give rise to discussions concerning the course of action to be taken. The clinical diagnosis is difficult and in practically all the cases described in the literature, the curative operation was performed without the diagnosis having been established pre-operatively. This was also true in our case, in which the diagnosis was not confirmed until the final histopathological examination despite the fact that fine needle biopsy with an ensuing cytological examination

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had been performed. If the diagnosis is suspected, an examination of freeze-sectioned material during the operation should be a possible means of providing the operator with valuable information at the time of the extirpation. Neither in our material nor in any other case described in the literature has this possibility been utilized.

The treatment of the condition is always surgical. The type and size of the intervention is extremely variable in the literature. Simple extirpations have been performed in many cases with good results and freedom from recurrences, while Walker and Resler (1963) consider that craniotomy should always precede other operative procedures. Like many other authors, we would stress the benign nature of the nasal glioma. Although recurrences have been reported, we have not been able to find a single case in the literature where the patient died as a result of invasive growth or metastases. We are therefore of the opinion that the surgical procedures should be relatively conservative and that mutilating, or grossly disfiguring operations should be avoided. In our case it is fairly certain that the operation was not radical. Nevertheless, a period of observation of four years has shown that the remaining glioma tissue has grown more slowly than the other healthy tissue. Viewed in conjunction with other case descriptions in the literature, this supports our opinion that the primary procedure must not necessarily be a radical operation. Thus, expectant treatment and surgery in the event of recurrences should be preferable to very extensive primary procedures. Regardless of the size of the operation, careful post-operative observation is required owing to the possibility of meningo-encephalitis.

Summary

A case of nasal glioma is presented along with a brief review of the aetiology, structure and diagnostic possibilities as published in earlier surveys. Considering the extremely benign nature of this type of tumour, we are of the opinion that the surgical measures may be relatively conservative and that mutilating, or grossly disfiguring, procedures should be avoided.

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