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TREATMENT OF PINEALOMAS BY STEREOTAXIC RADIATION SURGERY

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Tumours in the pineal region have always posed therapeutic problems when radical excision has been attempted. Experiences from various types of surgery in 140 cases (1926—1962) were published by OLIVECRONA (1967), with an overall mortality as high as 50 per cent. During the last ten years, more conservative modes of treatment have been used, including shunting procedures and radiation therapy.

At this department, stereotaxic radiation surgery has been applied to five cases of tumour in the pineal region with rewarding preliminary results in all cases, as a matter of course without complications or mortality. Three of these had comparatively small tumours, not causing much deformity of the CSF pathways. Furthermore, in these three cases, stereotaxic needle biopsy before the irradiation had given only scanty material which was not conclusive for diagnosis. The remaining two cases were selected for presentation, as in each of these, a large tumour was present occluding the CSF passage, the stereotaxic biopsy was conclusive and the cytologic appearance that of a pineocytoma, and a thorough follow-up including pneumography was performed, giving evidence of a considerable shrinkage of the lesion.

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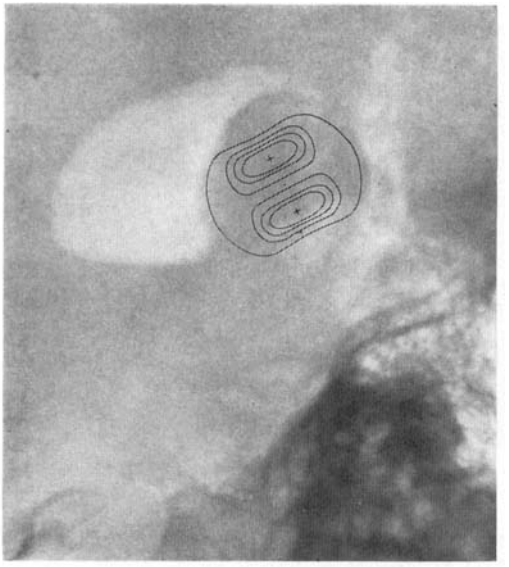
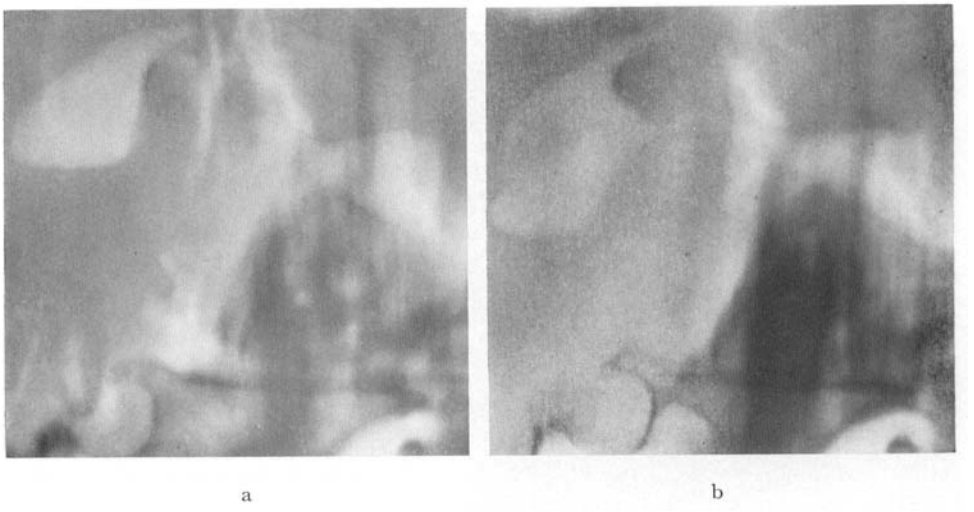


Fig. 1. Case 1. Preoperative encephalography. Deformation of the posterior part of the third ventricle by a rounded pinealoma. Curves of the dose distribution used in this case are superimposed, indicating 80, 60, 40 and 20 per cent isodose levels in relation to the dose at the four target points, two of which are indicated by crosses in this figure.

Method. The method of treatment has been fully described previously (LEKSELL 1971, BACKLUND et coll. 1972) and will only be briefly mentioned here. The procedure is performed by cross firing of the tumour by narrow beams from 179 small ^{60}Co-sources, distributed over a spherical sector. The beams are radially directed towards the centre of this sector, which is aligned with the stereotaxically predetermined tumour target point (or points) during the irradiation procedure. The apparatus (manufactured by AB Motala Verkstad, Motala, Sweden) was primarily developed for functional neurosurgery. Because of this, the system of apertures for the individual beams and the spatial distribution of the ^{60}Co sources were designed to give disc-shaped dose distributions. For tumour treatment, such a dose distribution may be too small and not of optimal shape. To overcome this and in order to align the irradiated volume to the shape and size of the tumour, a number of targets in a predetermined spatial pattern are used (Fig. 1). As may be seen from the figure, the dose edge gradient is very steep, thus permitting large doses to be given to the target area while negligible doses are delivered to the surrounding tissues. The irradiation is given in single doses and the dose rate at present permits the delivery of 1 000 rad to the target in the centre of a human head in about 10 minutes.

Case reports

Case 1. A 13-year-old girl was admitted in July 1969 with one month's history of attacks of headache, accompanied by nausea and vomiting. Bilateral papilloedema with 2 diopters' protrusion was confirmed on admission. At encephalography a deformation of the posterior



a b

Fig. 2. Case 1. Encephalography (a) before and (b) one year after radiation surgery. In (b), the tumour has diminished in size almost to that of a normal pineal gland and the aqueduct appears normal.

part of the third ventricle, indicating a rounded pinealoma 2 cm in size was demonstrated (Figs 1, 2 a). The aqueduct was displaced and partially obstructed, and marked ventricular dilatation was present (Fig. 3 a). No tumour vessels were seen at vertebral angiography. A stereotaxic aspiration biopsy of the tumour was performed and the cytologic appearance indicated a pineocytoma. The tumour was treated by stereotaxic radiation surgery. Four targets were chosen to given a dose distribution as optimal as possible (Fig. 1). A dose of 5 000 rad was delivered to each target, hardly any part of the tumour receiving less than 500 rad. The operation was performed under local anaesthesia. Postoperatively, the papilloedema disappeared rapidly and two weeks later no protrusion could be seen. Simultaneously, the symptoms and signs of raised intracranial pressure were relieved. At encephalography one year later, the appearance of the tumour region was almost normal (Fig. 2 b) and the lateral ventricles of ordinary size and shape (Fig. 3 b). On re-examination three years after the treatment, the patient was perfectly healthy and the ocular fundi were normal. Another encephalography revealed no obvious further change, although the pineal lesion was possibly still smaller.

Case 2. A 21-year-old man was admitted in February 1972 with a history of delayed puberty and diplopia, upward gaze disturbances, headache and polyuria for six months. A previous encephalography had revealed a hydrocephalus, apparently caused by a rounded expanding lesion about 2.5 cm in diameter, partially occluding the aqueduct and causing dislocation of the third ventricle (Fig. 4 a). On admission, panhypopituitarism was also established and the patient was subjected to full substitution. By stereotaxic biopsy the lesion was found to be a pineocytoma. In April 1972 the tumour was treated by stereotaxic radiation surgery. Four targets were chosen, with a distribution similar to that used in Case 1. The radiation dose to each target was 5 000 rad. One week after the treatment, papill-

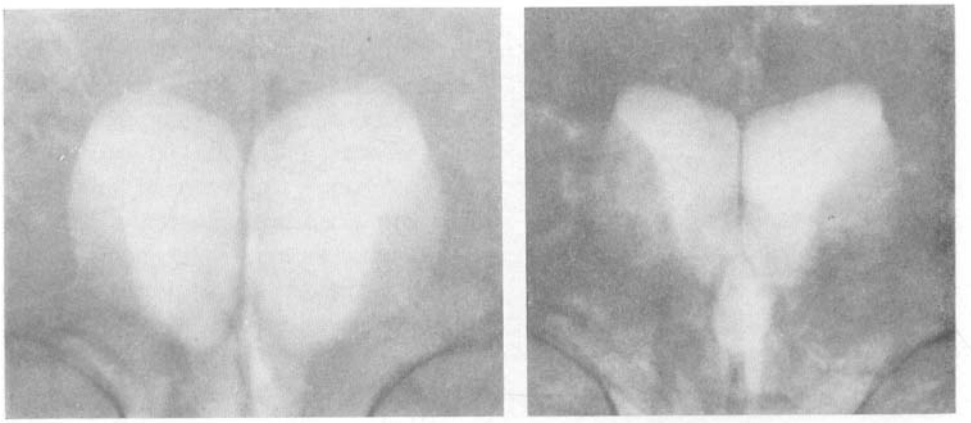


Fig. 3. Case 1. The ventricular system before (a) and one year after the treatment (b). The width of the lateral ventricles is less in (b), whereas the third ventricle is still somewhat widened.

oedema and symptoms of increasing intracranial pressure appeared. This was assumed to be due to occlusion of the aqueduct caused by oedema of the tumour, induced by irradiation. A ventriculo-atrial shunt ad modum Pudenz was inserted. The ventricular pressure was found to be considerably raised. After the shunt operation a rapid improvement was recorded. One month later, the shunt was found to be working inefficiently; nevertheless the patient was still improving. Four months after the radiation surgery the papilloedema and the diplopia had disappeared, but a slight weakness of upward gaze remained. At lumbar encephalography only the basal cisterns and the fourth ventricle could be filled; ventriculography revealed that the tumour had disappeared but no passage of air through the aqueduct could be demonstrated. This was considered to be due to a localized angulation of the aqueduct in the vicinity of the irradiated area, possibly resulting from the tumour shrinkage. A complete obstruction of the aqueduct was however not considered likely because of clinical evidence of adequate passage of CSF, as the papilloedema and subjective symptoms of intracranial pressure subsided in spite of the increasing malfunction of the shunt. The patient was subsequently transferred in very good condition for further evaluation of his hormonal state. A recent neurologic examination failed to reveal any defects apart from a slight weakness of upward gaze (March 1973).

Discussion

In spite of modern techniques, the surgical extirpation of pinealomas is still a problem and even in recent publications high mortality rates are reported. Suzuki & Iwabuchi (1965), in their presentation of 19 cases, gave an account of a 37 per cent operative mortality. In 13 cases, a complete removal could be performed, but in the remainder (32 per cent), the removal was less than total

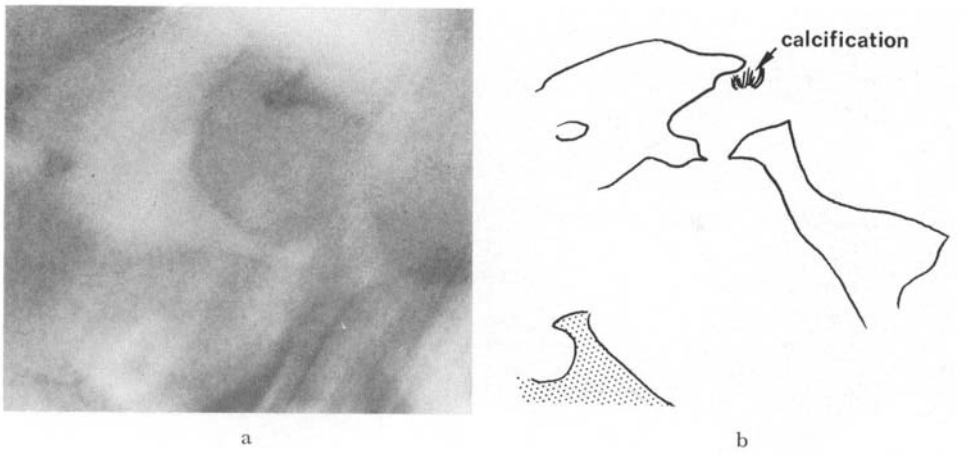


Fig. 4. Case 2. a) Encephalography before treatment. The tumour is indenting the third ventricle and causing considerable antero-inferior displacement of the aqueduct and fourth ventricle. b) Composite drawing from encephalography and ventriculography four months after radiation surgery. The shape of the third ventricle is almost normal. The fourth ventricle and the aqueduct were outlined by lumbar air injection.

to a varying degree. Of the lesions that were removed completely, four were cystic. In the majority of the successful cases, shunt procedures and postoperative irradiation were also performed.

In a series of 14 cases treated after 1950 with modern surgical techniques, POPPEN & MARINO (1968) reported only two fatal cases. However, complete removal was attempted in three cases only and, furthermore, the two fatal cases belonged to this group. These authors conclude: 'Even with the several techniques now available, the postoperative mortality rate is still high', and a conservative attitude was advocated.

Recently, 6 cases of lesions in the pineal region were reported by STEIN (1971), including one cystic astrocytoma, one epidermoid and one obscure case with arachnoidal cysts in the quadrigeminal region. Subtotal removal was performed in two cases, another two were submitted to biopsy and in the remaining two only cyst aspiration and exploration plus a shunting procedure were performed, respectively. No immediate postoperative mortality was reported, but the astrocytoma patient died one year after surgery.

A further six cases were reported by JAMIESON (1971), obviously constituting selected surviving cases from a larger unpublished series. The histologic grouping of these six cases was: atypical teratoma (3), pineocytoma (1), undifferentiated cyst (1), and normal pineal gland? (1). A fairly radical removal was achieved but in three cases gaze disturbances and other preoperative signs remained

unchanged after operation. In one case, it was necessary to make an occipital lobe resection, which caused a permanent hemianopia.

In general, those neurosurgeons who have made use of more conservative measures in pinealoma treatment report better clinical results.

CUMMINS et coll. (1960) assessed the value of radiation therapy in 37 cases of tumour of the third ventricle (22) and pinealomas (15). The operative mortality reported after direct surgical approach for excision was 34 per cent. The patients who were exposed to less radical surgery and were irradiated postoperatively had a 61 per cent five-year survival rate. The authors are definite in their conclusion that the preferred method is radiation alone or combined with shunting, if obstructive hydrocephalus is present. In a paper by SMITH (1961), presenting a large series including cases previously reported by RAND & LEMMEN (1953), a similar conclusion was drawn.

It is interesting to note the change of opinion of SUZUKI in his presentation of a second pinealoma series (SUZUKI & HORI 1969), in which he advocated shunting and radiation therapy. The authors quote several Japanese colleagues with similar experiences. One fatal case in this latter series however indicates the potential risks with high-voltage irradiation to targets close to the brain stem. Fortunately, disastrous courses after radiation therapy in similar cases are infrequent, but illustrate the main drawbacks of this type of treatment (GHATAK & WHITE 1969, HOLDORFF & SCHIFFTER 1971). Such disadvantages can be avoided and the risks reduced to a minimum by stereotaxic radiation surgery, whereby the radiation is delivered almost exclusively to the tumour, making possible the selective irradiation of neoplasms in the pineal region, and at ectopic sites, separately.

In order to utilize maximally the intrinsic precision of this type of treatment, a precise radiologic mapping of the tumour is a prerequisite. Encephalography and vertebral angiography make such an accurate mapping possible and in many cases also permit a determination of the character of the tumour (LÖFGREN 1958, GREITZ 1972).

It is often stated that any course of radiation therapy should be based upon histologic verification of the lesion to be irradiated. In radiation surgery on the other hand this principle is less critical as the purpose is complete necrosis of the irradiated area. Nevertheless, stereotaxic biopsy was performed in the two cases presented, for two main reasons: (1) Cystic lesions should not be treated with closed radiation surgery, but may be suitable objects for intracystic isotope treatment (BACKLUND et coll.). (2) Histologic classification certainly influences the determination of the dose level, as radiation doses higher than the minimum dose necessary for necrosis are pointless.

Fractionated radiation doses of 3 500 to 6 200 rad (SUZUKI & HORI), 4 000 to 5 000 rad (CUMMINS et coll.) and 3 500 to 4 200 rad (ENNUYER et coll. 1956) delivered with a conventional technique have proved to be fully effective in preventing continued growth of pineal tumors. As the biologic effect produced by a single dose is greater than that produced by the same dose fractionated, single doses of 5 000 rad have been considered sufficient to cause obliteration of the two pinealomas presented. A considerable shrinkage of the tumours was evident although complete disappearance of the lesions could not be demonstrated. The radiologic findings after the treatment may however be explained by the fact that tumour remnants after irradiation may consist of a non-vital mass of debris (SUZUKI & HORI).

With the introduction of stereotaxic radiation surgery, new possibilities are offered for treating other benign and circumscribed but relatively inaccessible lesions of the brain, such as craniopharyngiomas and other pituitary tumours as well as acoustic neurinomas.

This type of therapy has been used in a series of such cases since 1968 with encouraging results. In craniopharyngiomas and pituitary adenomas not only a permanent obliteration and shrinkage of the tumour has been demonstrated but also, in many cases, improvement of the endocrine state of the patients (BACKLUND 1973, 1974). In the field of acoustic neurinoma treatment, it has been possible, in a small series, to irradiate selectively tumour remnants in the orifice of the internal meatus after conventional surgery. The experiences hitherto indicate that radiation doses sufficiently great to cause shrinkage of the tumour remnants can be delivered without simultaneously being noxious to the facial nerve. This is supported by the general conception in radiation biology that axons are very resistant to irradiation (cf. RUBIN & CASARETT 1968). For small acoustic tumours, radiation surgery has been used as primary treatment and one case has been reported (LEKSELL 1971).

With the method of treatment presented, it has been possible to obtain striking therapeutic effects both as regards the tumours and the clinical condition of the patients. The clinical course in these two patients furthermore was very smooth, contrasting markedly with the often very dramatic postoperative course after radical surgery; stereotaxic radiation surgery is therefore considered the method of choice in cases of pinealoma.

SUMMARY

Two cases of pineocytoma, treated by stereotaxically directed gamma radiation from 179 ^{60}Co-sources are presented. Before treatment, the tumours were classified by stereotaxic fine needle biopsy. In both cases, signs and symptoms of raised intracranial pressure

disappeared soon after the irradiation. Encephalography revealed almost complete disappearance of both lesions. The advantages of this type of pinealoma treatment as well as the general principles for stereotaxic radiation surgery in cases of small, benign, inaccessible intracranial tumours are discussed.

ZUSAMMENFASSUNG

Zwei Fälle mit einem Pineocytom, die mit stereotaktisch gerichteter Gamma Bestrahlung durch 179 ^{60}Co Strahlenquellen behandelt worden waren, werden vorgestellt. Vor der Behandlung waren die Tumoren mittels stereotaktischer Feinnadelbiopsie klassifiziert worden. In beiden Fällen verschwanden die Zeichen und Symptome eines gesteigerten intrakranialen Drucks rasch nach der Bestrahlung. Die Encephalographie ergab ein beinahe vollständiges Verschwinden beider Läsionen. Der Fall der Behandlung eines Pinealoms sowie die generellen Prinzipien der stereotaktischen Strahlenchirurgie bei Fällen mit kleinen, beningnen, nicht erreichbaren intrakranialen Tumoren werden diskutiert.

RÉSUMÉ

Présentation de deux cas de pinéocytome, traités par la radiation gamma de 179 sources de ^{60}Co. Avant le traitement les tumeurs ont été classées par biopsie stéréotaxique avec une aiguille fine. Dans les deux cas les signes physiques et les signes fonctionnels d'hypertension intracrânienne ont disparu peu de temps après l'irradiation. L'encéphalographie a montré une disparition presque complète des deux lésions. Les auteurs examinent le cas du traitement des pinéalomes ainsi que les principes généraux de la radiochirurgie stéréotaxique dans les cas de petites tumeurs intracrâniennes bénignes inaccessibles.

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