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Radiation therapy in the management of chemodectomas in Nigeria

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Summary

Chemodectomas constitute a very rare group of tumours. They have been described at various sites in the body and their management, particularly the glomus jugulare tumours is of interest, as surgery in some cases may be hazardous and possibly fatal as a result of severe haemorrhage associated with these tumours.

Radiotherapy has been advocated as the treatment of choice particularly for the glomus jugulare tumour.

Our experience in the management of this rare group of tumours seen at the only Radiotherapy Unit in Nigeria between 1975 and 1979 is analysed. Radiation therapy techniques, doses given and complications noticed are analysed.

There was complete control of the disease in four out of four (100%) of the cases at 1 year and three out of four (75%) at 3 years. Results from other reports in the literature are reviewed.

Résumé

Les chemodectomas constituent un group rare des tumeurs. On les décrit dans plusieurs parties du corps et leur controle est intéréssant, notamment celui des tumeurs glomus jugulaire, étant donné que la chirurgie pourrait être hasardeux et peut-être fatale par suite d'une grosse hémorragie associée à ces tumeurs.

On a préconisé la radiotherapie le traitement du choix, notamment pour les tumeurs glomus jugulaire.

Voilà l'analyse de notre expérience dans le contrôle de ce groups rares des tumeurs vu dans la seule bloc radiothérapie au Nigeria entre 1975 et 1979.

Les techniques de la thérapie de radiation, dosages données, et les complications remarquées sont tous analysés.

Il y avait un contrôle total de la maladie dans quatre sur quatre (100%) des cas en 1 an et trois sur quatre (75%) en 3 ans. Des résultats des autres rapports en documentation sont passés en revue.

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Introduction

Chemodectomas constitute a very rare group of tumours (Berk, 1961; Parisier & Sinclair, 1968). They arise from the non-chromaffin paraganglia chemoreceptor cells. These tumours have been described in various sites such as the dome of the jugular bulb (glomus jugulare) (Bradshaw, 1961; Hawkins, 1961; Palacois, 1970), the carotid body (Bosniak *et al.*, 1964) aortic body, the vagus nerve (Kircher, 1967), ciliary ganglion of the orbit, nose, (Parisier & Sinclair, 1968), lung (Ichinose, Hewitt & Drapinas, 1971).

Surgical management, even where possible, is often hazardous because of the heavy haemorrhage that may result from these highly vascular tumours.

Radiotherapy is the treatment of choice particularly for the glomus jugulare tumours.

The experience from the radiotherapeutic management of the cases seen between 1975 and 1979 at the Radiotherapy Unit of the Lagos University Teaching Hospital is presented.

Materials and methods

The Lagos University Teaching Hospital is at present the only centre in Nigeria with facilities for radiation therapy.

The records of all cases with chemodectoma treated at the Radiotherapy Unit between 1975 and 1979 are analysed with emphasis on their mode of presentation, investigations and the details of radiotherapy. The complications and their follow-up records were analysed. The tumour was considered to be controlled if at 1 year after radiotherapy, there was no increase in size or progression of symptons. The 1-year period was chosen because of the immense local problem with patients, follow-up where the rate of default is high.

Results

The salient clinical features at presentation are as shown in Table 1. All four patients analysed underwent the following investigations to confirm diagnosis: (i) biopsy, and (ii) carotid angiograms.

One of the patients (No. 142160) had a very severe haemorrhage following her biopsy and required transfusion thrice subsequently. Other routine baseline investigations, such as haemogram, serum biochemistry, skull X-rays, etc., were essentially normal in all the patients.

Figure 1 shows the external carotid angiography for one of the patients with glomus tumour extending upward into the petrous area on the right side. The tumour mass and its blood supply is shown well above the carotid bifurcation. The mass is pushing the internal carotid segment anteriorly. Its beaded appearances are also noted.

Treatment

Table 2 shows the details of radiotherapy including the dose fractionation schedule. Some complications observed are also listed.

Radiotherapy plan

One patient was treated with the Orthovoltage

Table 1. Various signs and symptoms at presentation

Patient No.	Age (years)	Sex	Signs and symptoms
201509	39	F	Noise (L) ear — 8 years (L) facial palsy Mass (L) ear.
186636	45	F	Painful boil-like swelling (R) ear; partial deafness (R) ear; tinitus, vertigo } 3 years Recurrent bleeding (R) ear — 2 years (R) 5th and 7th nerve palsies
142160	45	F	Pulsatile swelling (R) retroauricular region — 3 years. Bleeding from swelling requiring transfusion 3 times. Mass reddish and almost completely blocking EAM* (R) 7th nerve palsy.
092969	46	F	Pain (R) ear — 4 years. Pulsatile polypoidal growth EAM* (R) 7th nerve palsy.

*EAM. = external auditory meatus.

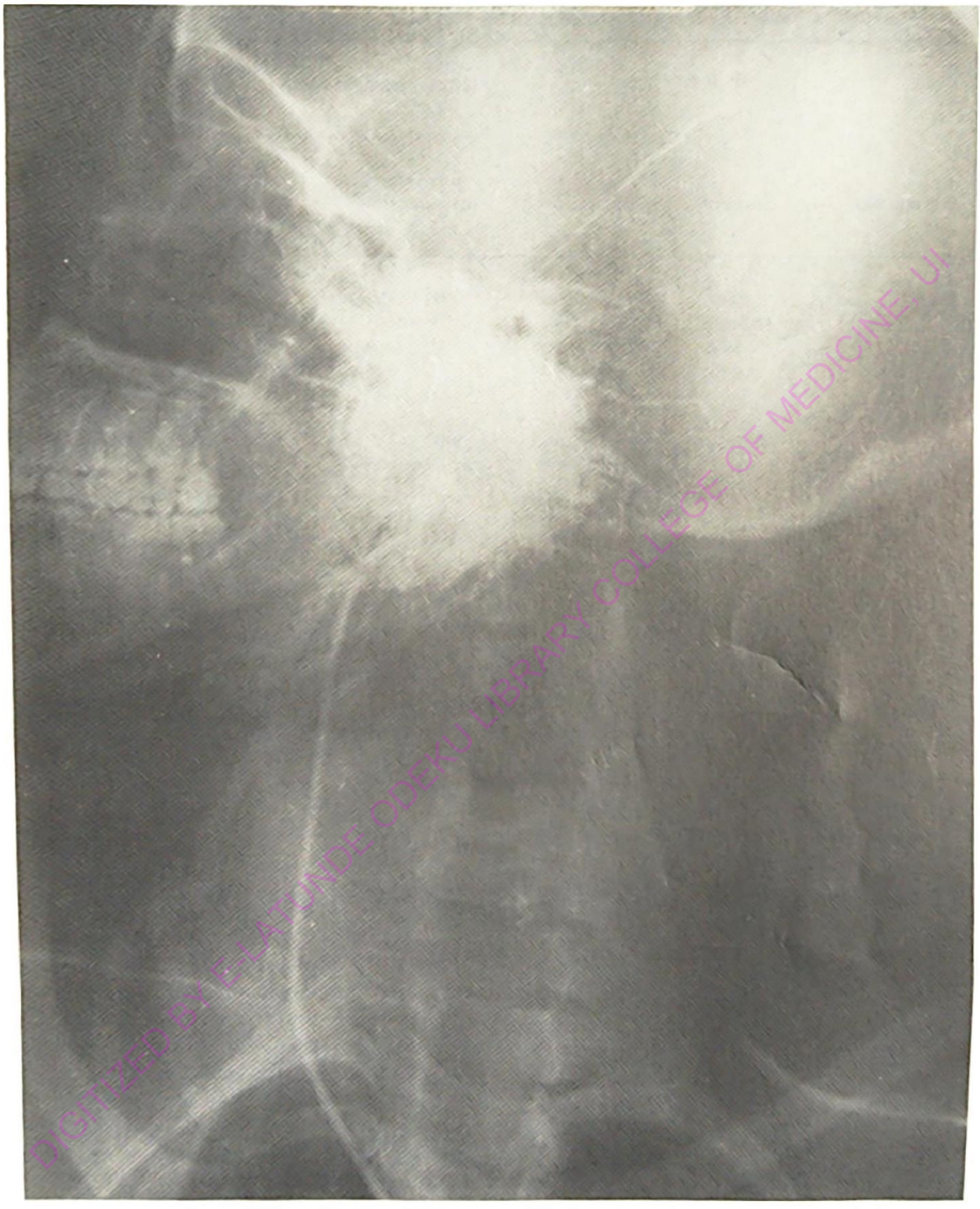

Fig. 1. A typical carotid anagram

machine (300 kV), whilst all others had treatment on the Cobalt-60 machine using a wedged pair of fields as seen in Fig. 2 with a lateral wedged field with its central axis along the external auditory meatus, and a posterior oblique wedged field with the axis along the direction of the petrous temporal bone. The exit beam is tilted to avoid the eye on the

Table 2. Details of management received and complications

Patient No.	Surgery	Radiotherapy details: Technique	Radiotherapy details: Dose/time fractionation	Complications
201509	Partial excision biopsy	Single direct field on Orthovoltage machine (300 kV)	5000 cGY in 5 weeks	Severe skin desquamation discharge (L) ear. Radiation fibrosis (L) Petrous bone
186636	Biopsy	Megavoltage wedge pair on Cobalt-60 machine	5000 cGY in 5 weeks	Oedama (R) face
142160	Biopsy	Megavoltage wedge pair on Cobalt-60 machine	4500 cGY in 4 weeks	Purulent discharge (R) ear. Triamus two further episodes of severe haemorrhage before completion of XRT
090969	Biopsy	Megavoltage wedge pair on Colbalt-60 machine	4500 cGY in 4 weeks	Dry skin; desquamation; Crusty discharge (R) EAM.

contralateral side. Field sizes varied between 6 × 6–7 × 7cm. The skin-sparing effect and the increased depth dose from the Cobalt beam make it more superior to the Orthovoltage beam. Another advantage is the absence of defferential bone absorption which reduces the risk of bone necrosis.

Assessment of control

The disease was classified as controlled if 1 year after treatment there is no evidence of increase in size or progression of symptoms.

Patient follow-up presents a lot of problems generally in Nigeria as most are in default.

Table 3 shows the state of the disease in the patients as at their last follow-up. At the time of this review, all the patients except one are lost to follow-up.

Discussion

All the cases of chemodectoma described in this paper arose from the glomus body, a small chemoreceptor organ situated in the wall of the jugular bulb. All the cases occurred in middle-aged females, between 39 and 46 years, each presenting with a mass in the external auditory meatus. The tumour is visible in the (R) ear in three out of the four cases. Miller (1962) showed a similar preponderance of (R) ear tumours whilst Moore *et al.* (1973) showed involvement of (L) ear in 70% of the thirty-three cases reported.

Various reports have shown involvement of cranial nerves particularly the 5th, 7th, 9th, 10th, 11th and 12th nerves, most of which leave the skull through the foramina near the jugular foramen and are therefore easily involved by the tumour. This series only showed a uniform

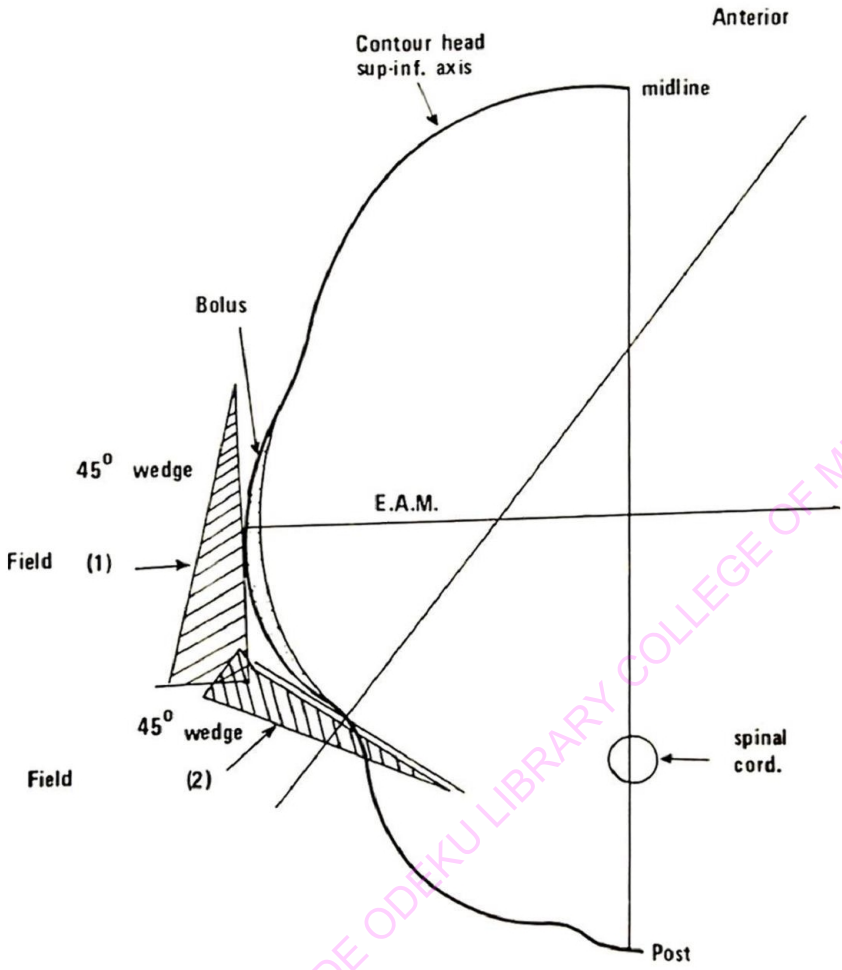

Fig. 2. Radiotherapy fields used in treatment planning on Cobalt-60 machine.

Table 3. State of disease as at last follow-up

Patient No.	State of disease/ symptoms	Duration of control at last follow up (years)
201509	Controlled	3
186636	Controlled	2
142160	Controlled	7
090969	Controlled	3

involvement of the 7th nerve in all cases and the 5th nerve in one other patient. It is the facial palsy that prompted two of these patients to seek medical help. Miller (1962) suggested that when only the facial nerve is involved, the tumour is restricted to the vicinity of the jugular bulb whilst involvement of other nerves suggests extension into the posterior fossa.

The long duration of symptoms in these patients is noteworthy, with a mean of 4 years (range 3–8 years.) Longer periods have been described thus showing that the tumour is slow growing and locally invasive.

Routine biopsy of these tumours is not advisable as this may lead to torrential and possibly fatal haemorrhage. An angiogram is very essential for diagnosis and also for pre-operative assessment or for radiation-treatment planning.

Embolization has been tried and found useful particularly before surgery as it may reduce the severity of haemorrhage (Ruggierd, 1976; Picard, Frost & Roland, 1976).

Radiotherapy, however, is the treatment of choice for the glomus jugulare tumours particularly as the risk of haemorrhage from surgery is avoided, morbidity is less and there is a high rate of control of symptoms.

The optimum dose of radiation to be given has been the subject of controversy, and doses ranging between 3500–4500 and 5000–6000 cGy have been used.

Patients who received over 5000 cGy in 5 weeks have been reported to have increased incidence of complication particularly brain necrosis. Kim, Elkon and Lim (1980), in a literature review of over 200 patients found a 2% recurrence rate in patients who had 4000 cGy in 4 weeks or higher whilst 22% recurred amongst those who had less than 4000 cGy. There was no evidence of brain necrosis in our series and there was adequate control of the disease in four out of four (100%) of our patients at 1 year, and three out of four (75%) at 3 years, although none of them showed any recovery of their facial palsy.

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