CARCINOMA OF THE MIDDLE EAR*

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SUMMARY

Five cases of advanced carcinoma of the middle ear and mastoid are presented. One was a papillary adenocarcinoma of parotid gland origin; 1 was an anaplastic carcinoma; 3 were well-differentiated carcinomas. The diagnosis and assessment of this condition is reviewed. Radiotherapy followed by petrosectomy in operable cases is recommended as the treatment of choice.

Carcinoma of the middle ear and mastoid is a rare condition. The proportion of this disease to all other pathological conditions of the ear has been reported by Towson and Shofstall[1] as 1:5 000 to 1:7 000; by Mattick and Mattick[2] as 1:7 000; by Furstenberg[3] as 1:20 000; and by Schall[4] as 1:6 000. This report documents my personal experience of 5 cases occurring in South African Bantu. These patients were treated over a period of 3 years at Baragwanath Hospital where 85 000 cases are admitted annually.

Carcinoma may arise primarily in the middle-ear cleft, or may spread there from surrounding structures, viz. the auricle, external auditory canal or parotid gland. Mattick and Mattick[2] reported that only 1·5% of all ear carcinomas originated primarily in the middle ear or mastoid. Lewis[5] reported 150 cases of ear cancer, of which 10 were primary squamous carcinomas of the middle ear, 2 were primary squamous carcinomas of the mastoid, and 2 were adenocarcinomas extending from the parotid gland. Primary carcinoma of the Eustachian tube has been reported by Passe[6] and by Siedentop and Jeantet.[7] The majority of cases reported are squamous carcinomas. Primary adenocarcinoma of the middle ear is extremely rare. Siedentop and Jeantet reported 3 cases and discussed 9 other cases in 1961.

TREATMENT

The treatment of carcinoma of the ear has undergone a change in recent years. For many years the standard treatment was radical mastoidectomy followed by radiotherapy. Lederman[8] advocated preliminary mastoidectomy as a means of defining the limits of the tumour before treatment with radiotherapy. Since then, the refinements in polytomography or multiplane tomography have made it possible to delineate the extent of the tumour very accurately. Boland and Patterson[9] suggest that preliminary mastoidectomy before radiotherapy may be frankly harmful. They reported 27 cases with 6 survivors over 5 years. In 5 of these 6 survivors there was no surgical operation other than biopsy. Ten patients had a mastoidectomy a few weeks before the commencement of radiotherapy, and all died with presumed recurrence. Boland[10] reports on 18 patients treated on the linear accelerator, of which 10 were alive and well after 5 years. There is substantial improvement in the results of radiotherapy when 4-MeV X-rays are used in a 3-field arrangement.

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Surgical Technique

While radiotherapy techniques have improved, fundamental improvements in the surgery of this area have also been achieved. Campbell *et al.*,[11] Ward *et al.*[12] and Parsons and Lewis[13] have pioneered the operation of total petrosectomy and indicated its feasibility for this disease. Incisions are planned to encircle the tumour and any areas of skin which are infiltrated or devitalized following irradiation. The incision extends down the sternomastoid muscle for 2.5 cm, and may be extended for a block dissection of the neck glands. The tumour is then circumvented by incision and reflecting the temporalis and sternomastoid muscles; the zygoma and masseter may be divided; the mandible may be disarticulated anteriorly to improve the exposure, or the ascending ramus may be divided. The parotid gland is dissected to expose the carotid artery, the jugular vein, the 9th, 10th, 11th and 12th cranial nerves and the sympathetic chain. The squamous temporal and mastoid are then drilled away to expose the sigmoid and petrosal sinuses and jugular bulb. The block of tissue including the tumour and petrous bone is then removed by rocking and fracturing the petrous bone lateral to the internal carotid artery. Retraction of the temporal lobe and cerebellum may be facilitated by withdrawing cerebrospinal fluid by lumbar puncture or by the intravenous injection of 30% urea in 10% invert-sugar solution. The internal carotid artery is then exposed by gently removing the overlying bone with a burr or bone-nibbling forceps. The artery is not ligated. The lateral sinus is frequently thrombosed and does not usually present a haemorrhagic problem. Venous bleeding may be controlled with packs of oxycell wool. Although the petrous pyramid may be removed entire, a piecemeal removal may be unavoidable if the bone is eroded or irradiated and breaks up, despite careful manipulation. If necessary, the apex of the petrous bone can be further exenterated with the drill. The dura is usually resistant to invasion, but in some cases it may be excised and a skin flap placed directly on the exposed arachnoid.

Allen[14] has extended the principles of this operation in the treatment of carcinoma of the parotid gland, or carcinoma of the ear which has invaded the parotid. The operation he describes involves neck dissection, mastoidectomy and complete removal of the mastoid tip and bony external meatal wall except for an anterior buttress. This procedure allows for total parotidectomy, providing access to all the extensions of the gland.

CASE REPORTS

Case 1. A 19-year-old Bantu female presented with a painful right ear and dizziness as her main complaints. On examination she was found to have a small mass behind the right pinna overlying the mastoid tip, and hard fixed lymph nodes were palpable in the right jugular chain. A fungating tumour was visible deep in the external auditory canal, associated with a purulent discharge. A history of chronic otorrhoea in that ear since childhood was elicited. The left ear was normal. Com-

plete paralysis of the following cranial nerves was discovered on the affected side: 5th, 6th, 7th (lower motor neurone lesion proximal to the geniculate ganglion), 8th (stone-deaf and spontaneous nystagmus to the left), 9th, 10th (right vocal cord paralysis), 11th (poor sternomastoid movement may have been due to fixation of the muscle by metastatic lymph node involvement), and 12th (wasting and fibrillation of the right side of the tongue). X-rays showed extensively pneumatized mastoids, with a huge destructive lesion of the middle ear destroying the jugular bulb and extending anteriorly to the petrous apex to involve the foramen ovale. Biopsy of the granulations in the external auditory canal showed the presence of an anaplastic carcinoma, the primary site of which could not be stated. Biopsy of the lymph nodes in the neck yielded a similar report. The tumour was too extensive for surgical treatment and she was treated with cobalt teletherapy, 6 000 rads. Some improvement in the pain was noted but 6 months later she was readmitted with severe back pain. A destructive lesion of the body and transverse process of L.5 and the right ala of the sacrum was noted. A further course of cobalt teletherapy was commenced. Three months later she developed severe pain in the hip. A pathologic fracture of the right trochanter and dislocation of the hip was noted. She died 10 months after the first admission to hospital.

Case 2. A 67-year-old Bantu male underwent a parotidectomy in 1960 for a papillary adenocarcinoma of the parotid gland. Postoperative radiotherapy was administered and a block dissection of the neck was carried out 2 years later. He was well for 7 years and then developed paralysis of the face and deafness on the affected side. There was no history of previous ear disease. At this stage he was referred to the Ear, Nose and Throat Department. A lower motor neurone paralysis of the right 7th nerve was present, the lesion being proximal to the geniculate ganglion. He was stone-deaf in the right ear and the tympanic membrane was congested and retracted, but intact. The parotid area and neck appeared to be free of recurrence, and no systemic metastases were discovered. Polytomography and conventional X-rays of the petrous bone showed erosion of the parotid tip, facial canal, tegmen tympani and jugular bulb. The cochlea and semicircular canals appeared normal. A mastoid exploration was carried out. The middle ear cleft was extensively eroded by tumour which had invaded the base of the skull from the region of the jugular foramen. The lateral sinus was thrombosed and invaded by tumour. The tegmen tympani was eroded, but the dura was not penetrated. The facial nerve was surrounded by tumour which had destroyed the mastoid tip and filled the tympanic cavity. A small eroded ossicular remnant was present in the granulations. The postauricular incision was extended downwards into the neck, and a wide exenteration of the tumour was accomplished, removing the tegmen tympani, sinus and dural plates, mastoid tip, jugular bulb and styloid process. The carotid artery was not ligated as we felt that the tumour could be adequately removed from the base of the skull with the exposure obtained. Histological examination of the tumour showed papillary adenocarcinoma, the features of which suggested a metastatic deposit. The pathologist suggested investigation of the thyroid and kidney—these proved normal. The devitalized skin failed to heal by first intention, but the wound granulated and closed in 6 weeks. A further course of cobalt teletherapy was then administered. He remains well and without recurrence 18 months after the operation (Figs. 1 and 2).

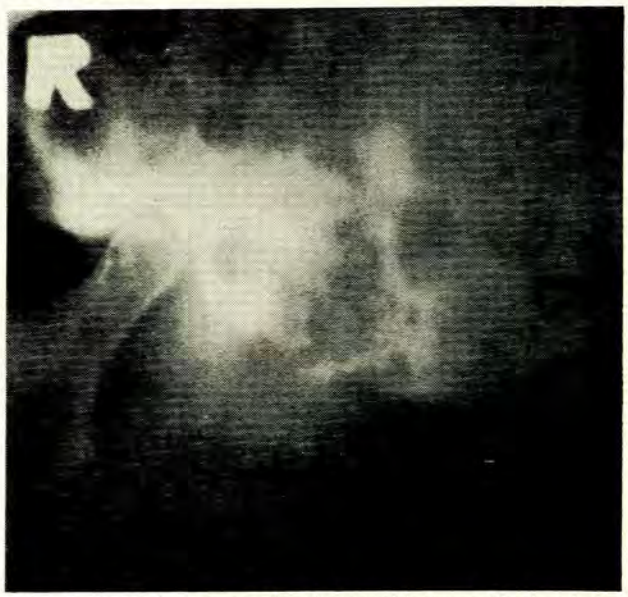

Fig. 1. Lateral tomograph of mastoid (case 2) showing erosion of tip, sinus plate, sinodural angle and facial canal.

Fig. 2. A-P tomograph of petrous bone (case 2) showing erosion of jugular bulb and destruction of ossicles.

Case 3. A 50-year-old Bantu male was referred to us with a huge friable fungating tumour of the right ear. The growth was 8 cm in diameter and no vestige of the external ear or canal was present. One year previously a biopsy taken from granulations in the external auditory canal had shown the presence of a squamous carcinoma and a course of radiotherapy (5 500 rads) had been administered. We were unable to trace the clinical details from the hospital where this occurred. On examination a

lower motor neuron right facial paralysis was present, the gag reflex was absent on that side and he was stone-deaf in the affected ear. No lymphadenopathy was noted. Biopsy of the cauliflower tumour showed the features of a well-differentiated squamous carcinoma. X-rays and polytomography of the petrous bone showed destruction of the mastoid tip, external auditory canal, tympanic cavity, ossicles, tegmen tympani and erosion of the cochlea and labyrinth. No metastases were detected. His haemoglobin was 10.5 g/100 ml and he was given a transfusion.

A wide excision of the tumour with a 2 cm margin of normal tissue was performed. The squamous temporal and occipital bones and zygoma were exposed. The sternomastoid and temporalis muscles were incised around the tumour. Removal of the tumour in a block together with the tegmen tympani and part of the squamous temporal, the sinus plate and mastoid tip was accomplished with the drill and gouges. The dura mater over the temporal lobe was not invaded. The lateral sinus was thrombosed and invaded by tumour. Granulations over the cerebellar dura mater were curetted away leaving an intact dura. At this stage the labyrinth and cochlea were exposed with residual tumour visible in the tympani cavity and solid angle. The petrous apex, including the cochlea and labyrinth were then exenterated with the drill. The carotid artery was not disturbed although a pre-operative angiogram had shown good cross-over circulation. The 9 cm cavity was then packed with B1PP. Skin grafting was not attempted. The wound granulated slowly and two months later a further course of 2 000 rads cobalt teletherapy was given. Five months later the wound was covered by smooth fibrous tissue with an epithelializing rim. The patient underwent this extensive procedure with remarkably little distress. He was quite well the day after operation, and complained only of headaches which lasted for 5 weeks. He is alive and well at the present time—1½ years after surgery (Figs. 3 and 4).

Fig. 3. Lateral tomograph of mastoid (case 3) showing erosion of tip, sinus plate, sinodural angle and facial ridge.

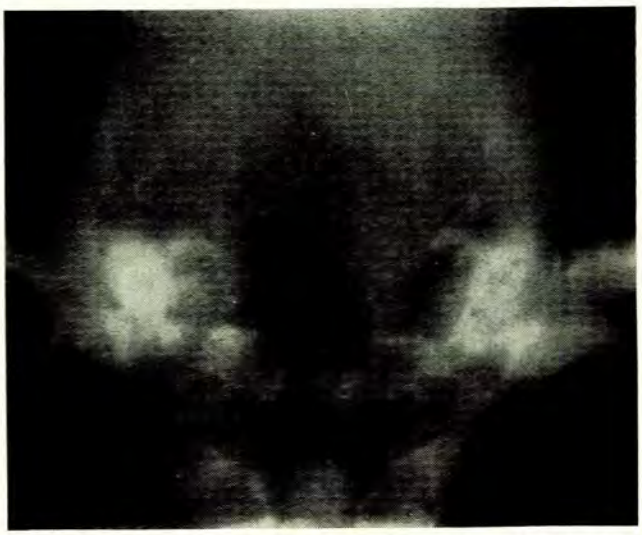

Fig. 4. A-P tomographs of petrous bone (case 3) showing destruction of external auditory canal, middle ear cleft, ossicles and facial canal.

Case 4. A 67-year-old Bantu female complained of dizziness, deafness and facial paralysis on the right side. These symptoms had commenced 6 months earlier but chronic otorrhoea on the right side had been present for years. On examination a firm swelling 2·5 cm in diameter was palpated anterior to the tip of the right mastoid. A granular polyp was visible in the external auditory canal. Complete paralysis of the 5th, 7th, 8th, 9th and 10th cranial nerves was observed. Biopsy of the polyp showed the features of a well-differentiated squamous carcinoma. X-rays of the temporal bone showed erosion of the mastoid tip, tegmen tympani, foramen lacerum, foramen ovale, foramen spinosum, jugular bulb, middle-ear cavity, semicircular canals and cochlea. The erosion extended to within 1·5 cm of the foramen magnum. She was treated with cobalt teletherapy (6 000 rads) and died of presumed local recurrence and cachexia 20 months after the first admission (Figs. 5 - 7).

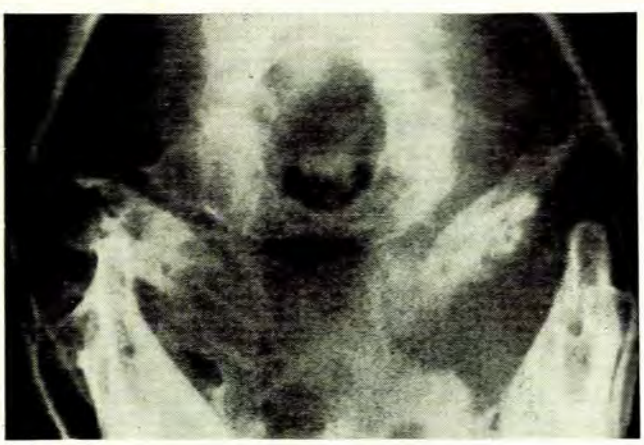

Fig. 5. Basal view of skull (case 4) showing gross destruction of petrous bone, erosion of foramen ovale, foramen spinosum and foramen lacerum.

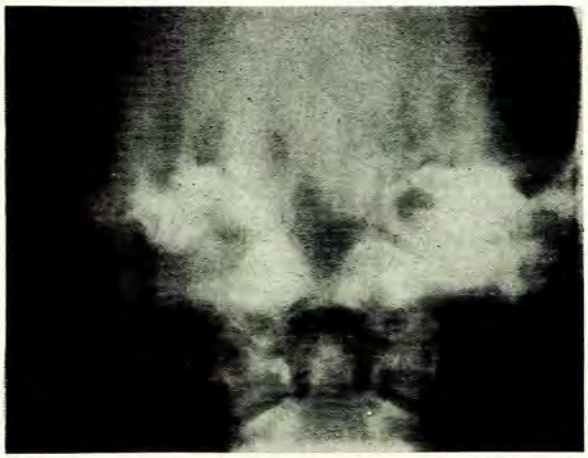

Fig. 6. A-P tomograph of petrous bone (case 4) showing destruction of external, middle and inner ear, jugular bulb and petrous apex.

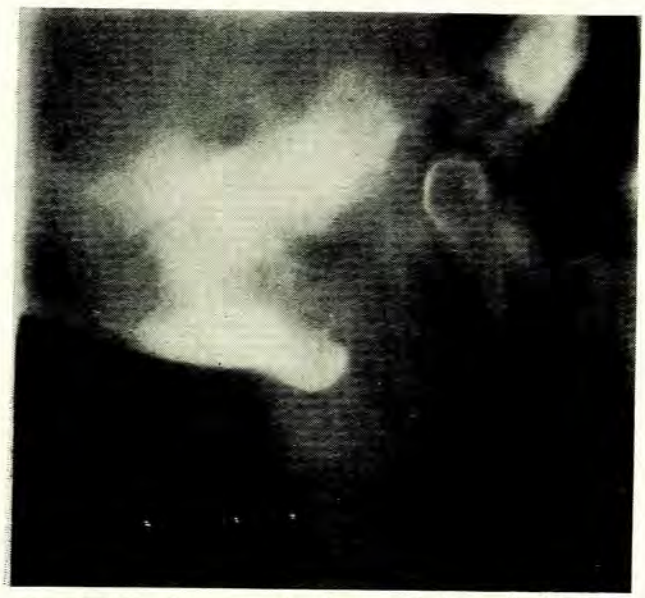

Fig. 7. Lateral tomograph of mastoid (case 4) showing erosion of middle ear cleft.

Case 5. A 42-year-old Bantu female complained of severe pain in the right ear and blood-stained otorrhoea. She had suffered from right purulent otorrhoea for years. On examination the right ear was stone-deaf and a lower motor neuron lesion of the facial nerve was present proximal to the geniculate ganglion. The corneal reflex was absent. The external auditory canal was filled with pink granulations. A small mass was palpable over the mastoid tip. A mass of hard lymph nodes was attached to the right sternomastoid muscle. Biopsy of the middle-ear granulations showed the features of well-differentiated squamous carcinoma. X-rays showed destruction of the mastoid tip and bowl, the middle-ear cavity and anterior surface of the petrous bone as far as the foramen ovale. The lesion was inoperable and she was treated with cobalt teletherapy (6 000 rads). The tumour melted away leaving a large cavity in the petrous bone which was visible through the external auditory meatus. She remains well 3 years after the conclusion of therapy.

DISCUSSION

In this series of 5 cases, 2 were well-differentiated squamous carcinoma arising in the middle ear or mastoid; one was an anaplastic carcinoma arising in the middle ear or mastoid; one was a papillary adenocarcinoma which spread from the parotid; and one was a well-differentiated squamous carcinoma involving the entire auricle and petrous bone, the site of origin being uncertain. In 2 of these cases metastases occurred, which most authors describe as being an unusual or late manifestation of the disease. There were 2 males and 3 females and their ages ranged from 19 to 67 years.

Early diagnosis is unlikely because of concealment and the fact that about half the cases had suffered from chronic otorrhoea for many years. Pain, deafness, bleeding from the ear and facial paralysis are common symptoms and occurred in all our cases. Involvement of the 9th, 10th, 11th and 12th cranial nerves indicates extension into the base of the petrous bone. Only 2 of our patients complained of vertigo. In all 5 cases a fungating tumour was visible in the depths of the external auditory canal. All 3 of the primary middle-ear tumours were associated with long-standing chronic suppurative otitis media.

We believe that carcinoma of the ear is best treated by a radical course of radiotherapy followed 5 weeks later by petrosectomy in operable cases. The question of operability is decided on polytomography in conjunction with basal views of the skull. Paralysis of the 9th, 10th and 12th cranial nerves does not specifically preclude surgery, but most of these cases will be found to have extensive spread anterior and medial to the petrous apex, and are therefore inoperable.

Two of our 5 cases have undergone petrosectomy following radiotherapy. Both recovered and are alive and well 18 months after the operation. The other 3 cases were considered to be inoperable and were treated with cobalt teletherapy alone. One survived 10 months, another 20 months and the third is alive after 3 years.

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