NASOPHARYNGEAL TUMOURS*

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The subject to be discussed is both wide and complex although this may not appear to be so and some difficulty has been experienced in deciding just how it should be treated. At first thought it is quite justifiable to question the necessity for accepting it as an entity worthy of separate consideration, but tumours as such are always of interest and usually of importance and I think that it can soon be shown that when they occur in the nasopharynx they create clinical pictures meriting some study.

There are two general methods of approach to the discussion, (1) the pathological, in which we accept the tumours which can and do occur and study how they may manifest their presence by the possible production of signs and symptoms, and, (2) the clinical, in which we find certain signs and symptoms and determine how they could be produced by tumour formation. The former tends to be theoretical and the latter a practical vindication, or otherwise, of the suggestion produced.

Let us begin with a few words about the location because it is by a disturbance of the form and functions of the structures in and around the nasopharynx that the tumours will declare their presence. Roughly speaking the nasopharynx lies behind the nostrils, below the base of the skull, in front of the vertebral column, between the ears and above the soft palate. The posterior openings of the nostrils form the greater part of its anterior boundary and the openings of the eustachian tubes lie in its lateral walls. The roof and posterior wall are continuous in a curved manner and the limits of the cavity as applied to the base of the skull are : posteriorly the pharyngeal tubercle in the basilar process of the occipital bone, then curving around forwards across the tip of the petrous bone, thus including the foramen lacerum, and to the medial pterygoid plate. Any tumour filling such cavity would cause nasal obstruction and resultant disturbance of speech and facies, also by interference with the eustachian tube a conductive deafness. Should the tumour be of the malignant type which spreads locally by penetration, then it would tend to follow any preformed anatomical pathway such as along the eustachian tube or up through the foramen lacerum. In the former case having passed out through the lateral muscular wall of the nasopharynx the tumour can spread in the deep retro-mandibular region just below the base of the skull and be likely to involve the muscles of the soft palate and/or their nerve

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supply and the third division of the trigeminal nerve. Spreading more posteriorly the tumour will reach the region of the jugular foramen and possibly involve the glossopharyngeal, vagus and accessory nerves or by even further spread, the hypoglossal nerve and sympathetic trunk.

Manifestation of such nerve involvement will be by associated rather distant pains and muscular palsies. Spreading upwards the tumour will penetrate the base of the skull through the foramen lacerum and come into association with the cavernous sinus. In this situation it may involve the oculomotor, trochlear and abducent nerves with more distant spread bringing it into association with the trigeminal nerve ganglion.

So much, quite briefly, for the location and now a word about the tumours. Broadly speaking they will be either simple or malignant and will thus differ in the way they can produce signs and symptoms. The simple tumour is merely a space-occupying lesion although of uncontrolled pertinacity. In the first place it will fill up the nasopharynx, but not stopping at that, will tend to expand the walls, always following as well, every natural path of expansion such as along the nostrils. Let us always remember that a mass of enlarged adenoids will behave very like this and produce the same signs and symptoms to begin with but it will stop at filling the available cavity and not stretch and distort the confining walls. The malignant tumour will, on the other hand, invade and penetrate the walls of the cavity without necessarily producing any great local mass. Now we encounter many different growth characteristics or one might say degrees of malignancy—some tumours tending to behave much as the simple neoplasm, others tending to spread into the local surrounding tissues, whilst others again tend to metastasize early through the lymph channels to the cervical lymph nodes and more distantly through the blood stream.

Now we have a general idea where our tumours are going to occur and how they produce signs and symptoms depending upon their growth characteristics. So we must proceed to a closer consideration of the actual tumours which do occur.

It is usual to include these tumours of congenital origin, the dermoids, teratomata and epignathi, but they can hardly be classed as real naso-pharyngeal tumours. They arise at the junction of mouth and pharynx and protrude as a rule from the mouth. They present no great diagnostic features apart from their actual presence, and treatment, if it is possible, will be by surgical removal. The dermoids are usually present at birth, fill the mouth from a mid-line origin and are often covered with hair. They are bi-dermal in origin being covered by skin but having perhaps bone and muscle inside. They may have an intra-cranial connection. The teratomata are more complex, being tri-dermal, appear early in fœtal development and tend to go on growing. The epignathi are parasite fœtuses and often show formed limbs.

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The Simple Tumours

Simple tumours may be of any type and are very rare, the only one of usual occurrence being the so-called fibroma. There has been a lot of confusion regarding this tumour by the inclusion of nasal polypi which hang through the posterior nasal choanae into the nasopharynx. The fibroma is quite a definite clinical entity, although doubt has been cast upon it as a pathological neoplasm, it being suggested as of inflammatory origin or even something akin to a keloid. Characteristically it is found in young males, is of varying rate of growth, and ceases to enlarge when the host reaches the age of twenty or so—hence the common additional name of juvenile fibroma. Many pathological labels have been applied, occasioned by the difference is histology found, but on the whole it remains that of a very vascular fibroma of varying growth rate to the extent in some cases of being active enough to be called a sarcoma, and behave as such. Usually the mass arises from a broad base in the region of the basi-sphenoid but as it enlarges to fill the nasopharynx and undergoes recurring superficial infections, it acquires secondary attachments and eventually may have very complex connections.

The signs and symptoms of such a tumour are as one would expect from a space occupying lesion : nasal obstruction with associated rhinophonia and dysphagia, conductive deafness due to eustachian tube obstruction and probably hæmorrhage due to surface infections, much the same as one might expect from enlarged adenoids although in this case the patient is older, even at that the case is sometimes only diagnosed by digital examination at operation for enlarged adenoids. Later, the tumour may grow and spread open the face, which associated with resultant exophthalmos, has been called " Frog face ".

Left untreated the lesion is said to run a fatal course in about three to six years, but if it does not form until the patient is older, say in the twenties, then growth may cease and it is said retrogression occurs. Treatment, as with any simple tumour which requires treatment, should be complete surgical removal, but unfortunately that is far from easy in this case. The locality is very difficult of access and owing to the great vascularity of the tumour, free and profuse hæmorrhage is likely to result from attempts at removal. Previous to the days of blood transfusion, many deaths occurred from this hæmorrhage. The present position is difficult to assess owing to the rarity of the lesion. It is to some extent radiosensitive, but will not, as a rule, undergo complete retrogression; for this reason irradiation tends to be used interstitially as an adjunct to other forms of treatment. To limit the vascularity and try to produce tumour necrosis, injections of sclerosing fluids, such as sodium morrhuate, have been made into its substance and with the same object in view, multiple punctures with a diathermy needle are used. On the whole these conservative measures are generally preferred to complete removal

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or attempts at such ; these are repeated frequently as needed and may cure, tide over to natural regression, or make removal possible. It should still, however, be kept in mind that removal is the ideal and only use other methods if it is definitely decided that such is impossible or too dangerous.

The Malignant Tumours

A whole variety of malignant neoplasms with varying growth characteristics occur in the nasopharynx and we will see how such will determine the signs and symptoms produced or vice-versa how we can assess the growth character of the neoplasm from the clinical picture found. The more simple type will behave as a space-occupying lesion, whereas the highly malignant type will probably metastasize before declaring its presence locally ; between these two extremes, we find the locally infiltrating lesion which is of such interest from an anatomical point of view. Thus right at the beginning we realize the importance of the so-called degree of malignancy, degree of differentiation or call it what you will. We must put together the anaplastic carcinomata, the sarcomata and the lympho-epitheliomata as quite different from the keratinizing squamous carcinomata.

The actual pathology of these malignant neoplasms in the nasopharynx has been the source of much discussion, but for present purposes they will be grouped according to their character of growth and spread and it is found that the anaplastic carcinomata and the sarcomata account for about four-fifths of the total, the remaining fifth being the keratinizing squamous carcinoma group.

Taking the latter first, we find them as infiltrating lesions which do not as a rule tend to grow to any great size locally, but can of course produce some manifestations by their local presence, such as little bleeding, maybe unilateral nasal obstruction and/or middle-ear deafness. They will tend to metastasize to the cervical lymph nodes only late and usually before that will have penetrated the pharyngeal wall to produce signs and symptoms by interference with nerves. These neoplasms tend to occur in the lateral and superior walls of the nasopharynx and when infiltrating will follow any anatomical path available.

Spreading laterally the neoplasm will follow the line of the eustachian tube and spread into the deep tissue spaces of the neck just below the base of the skull. Added to possible nasal obstruction, epistaxis and local pain, we will then find a conductive deafness and various combinations of muscular palsies depending upon the nerves involved in the region of the jugular foramen. Common manifestations of this spread are unilateral palatal palsy and laryngeal palsy, but any of the local nerves can be involved by direct local spread or at a lower level by lymph node metastases. Spreading upwards through the foramen lacerum the neoplasm now spreads as an intracranial lesion and manifests its presence

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by referred pain through the branches of the trigeminal nerve and diplopia as the result of eye muscle palsies.

Thus when we consider the lesion of the type of a keratinizing squamous carcinoma which tends to infiltrate the tissues, then we can find signs and symptoms produced as a result of such penetration outside of the confines of the nasopharynx. These are grouped as we might expect into two groups, one associated with palsies of the eye muscles and usually declaring their presence by diplopia, and the other associated with palsies of the pharyngeal and neck muscles, especially palatal and laryngeal palsies of one side. Always it must be remembered that other signs and symptoms of the neoplasm, especially cervical adenitis will probably be present as well, and also that many other lesions can produce similar clinical pictures.

The more malignant types of neoplasm such as the sarcoma, or anaplastic carcinoma are likely to metastasize to the cervical lymph nodes and thus declare their presence before they produce any great local damage, which of course they may do later. Also distant metastasis may occur.

So much for the viewpoint of accepting the presence of a nasopharyngeal tumour, noting the signs and symptoms which can be produced by its continued growth and dissemination conditioned by its growth characters, and how the pathology and clinical picture are correlated. Now we must change to the viewpoint of the clinician who is confronted with the clinical picture and must deduce the pathological state of affairs therefrom. It will be noted that the simple tumours produce such a clinical picture that they will be likely to be seen by an Ear, Nose and Throat Surgeon in the first place, whereas the malignant tumours are likely to be presented to the general surgeon, physician or ophthalmologist. When we realize that similar clinical pictures are produced, often more commonly, by lesions other than nasopharyngeal tumours, it will not be wondered that mistakes are common. The laryngologist often claims credit for the ultimate diagnosis of the primary tumour site, but it must be remembered that he was probably asked to see the case only because such was already suspected. The problem involved will be better appreciated by a statement of the condition found when eventually diagnosed as a malignant tumour of the nasopharynx. The signs and symptoms produced can be classified roughly under four headings as they are referable to, (1) the cervical lymph nodes, (2) the ears, (3) the naso-pharynx and (4) involvement of nerves. In the fully developed and late clinical picture, combinations of abnormalities will be found present as in the following table :—

(1) Cervical adenopathy in 75 per cent. of cases.
(2) Ear symptoms in 50 per cent of cases.
(3) Naso-pharyngeal symptoms in 50 per cent. of cases.
(4) Neurological disturbances in 40 per cent. of cases.

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With the eye and jugular foramen types about equal.

Going back into the history of the cases to determine what were the first signs and symptoms noted, it will be found that they were about as in the next table :—

Cervical adenopathy as a first symptom, 32 per cent. of cases.
Nasopharyngeal troubles as a first symptom, 30 per cent. of cases.
Ear troubles as a first symptom, 23 per cent. of cases.
Neurological disturbances as a first symptom, 15 per cent. of cases.

The duration of the first evidence of the trouble, until the real diagnosis was made, will be found to be over six months in about half of the cases seen. In consequence of this period when the correct diagnosis has yet to be made, it is not surprising to find that many of the cases have been treated under a wrong diagnosis. A survey shows that treatment is supplied on a symptomatic basis under the four main headings as shown in the next table before the correct diagnosis is arrived at.

(1) Cervical glands treated as such, 47 per cent.
(2) Nasal treatment provided, 25 per cent.
(3) Ear treatment provided, 25 per cent.
(4) Dental treatment provided (trigeminal nerve pain), 3 per cent.

Enough then to show the complexity of the picture and how in actual practice it has proven a great diagnostic puzzle, often wrongly interpreted. Of the many signs and symptoms which are produced it can only be suggested that in the presence of one such, at least a thought should be given to the possibility of a nasopharyngeal tumour as a cause, whilst a combination of several such, makes for greater certainty.

Aids to Diagnosis

As to the means of diagnosis available over and above the mere knowledge that the various signs and symptoms mentioned can be caused, among other things, by a nasopharyngeal tumour, there are the special means available to the laryngologist which enable visual examination of the nasopharynx. To this must be added digital examination under an anæsthetic. In those cases which present symptoms referable to the eye, an X-ray of the base of the skull may show enlargement by bone erosion of the foramen lacerum. Finally biopsy of the primary tumour or one of the neck glands is essential to complete the case before proceeding to prognosis and then treatment. Never to be neglected is a general examination for the possible presence of distant metastases, a point of importance, because available evidence suggests that such are present at the end in about a quarter of the cases.

As regards prognosis of the untreated cases, we know little except that as long as the diagnosis is correct, the end will be fatal, and the state of affairs can be explained to the patient, if necessary, before proceeding to treatment.

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Treatment

Even after our brief survey of the subject, it will be realized that surgery plays little, or no part, in the treatment of these malignant tumours of the nasopharynx. After all, diagnosis seems to depend upon spread and metastasis, by which time, excision is impossible. We are left then with radiotherapy, either high voltage X-ray or radium beam as the only hope of cure or palliation, and no more need be said about this, except as it effects prognosis. In other localities when excision of the neoplasm is possible, the prognosis is likely to be better in the keratinizing squamous carcinoma than in the anaplastic lesion and in consequence we tend to speak of the former as being less malignant. In contrast, however, the highly malignant anaplastic neoplasms and sarcomata are usually radiosensitive and hence in the nasopharynx have a relatively better prognosis. Even at that, the outlook is on the whole, bad.

Prognosis

After treatment by adequate radio-therapy in the best hands, the total five year cure rate is not likely to exceed 20 per cent. of all malignant cases. In cases where neurological signs are present, this figure falls to about 10 per cent. The presence of neck glands is so common that their presence does not alter the prognosis much, but in their absence, the cure rate rises to about 30 per cent. When there is no X-ray evidence of bone destruction in the base of the skull, the cure rate is about 25 per cent. but in the presence of such, it falls to just over 10 per cent.

As might be expected after the remarks regarding degree of malignancy and radiosensitivity, it is found that 30 per cent. or so of the sarcoma types can be cured whilst the carcinomas can only manage about 10 per cent. or less if well differentiated. The lympho-epitheliomata types which are especially radio-sensitive, produce a cure rate of nearly 40 per cent.

Palliation

If the cure rate of these cases is low, much can be done to alleviate symptoms. The most troublesome symptoms arise from neurological disturbances and fortunately these are produced in the first place by pressure rather than direct infiltration into the nerves so that irradiation can palliate.

Nearly 75 per cent. of cases can be made symptom-free but in three-quarters of them, relapse will occur and will do so within one year.

Termination

When death occurs and it usually takes an average of two years to do so, it is often as a result of intracranial invasion, but about a quarter of the fatal cases have distant metastases.

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Nasopharyngeal Tumours

Finale

These nasopharyngeal tumours are quite rare, yet they are always stimulating in that they illustrate so many principles of growth and symptom production that they form excellent teaching cases and always interest the student. Realization of the possibility of early diagnosis raises the hope that experience of such cases may with benefit be applied to other problems.

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