CARCINOMA OF THE LACRIMAL SAC*

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BOTH benign and malignant primary tumours of the lacrimal sac are uncommon. Penman and Wolff (1938) give an analysis of 64 cases, including one of their own, and since that date further cases, nine of which are detailed in Table I, have appeared in the literature.

TABLE I
ANALYSIS OF CASES OF PRIMARY TUMOURS OF THE LACRIMAL SAC
REPORTED BETWEEN 1938 AND 1950

Year	Authors	Sex	Age	Duration	Pathology	Treatment and Remarks
1939	McCool	M	54	6 months	Mixed-cell tumour of salivary type	Excision and radium implant. No recurrence after 10 months
1940	Spratt	M	66	5 years	Epidermoid carcinoma	Exenteration of orbit and x-ray therapy
1947	Florentin and Poirot	?	?	?	Naevocarcinoma	
1947	Carlevaro and Landoni	M	49	?	Reticulo-sarcoma	Generalized reticulo-sarcomatosis
1947	Rousseau	F	79	?	Epithelioma	Concurrent tumour of skin of temporo-malar area and lacrimal sac. Treated with radium. Good result after one year
1948	Vrabec	?	?	?	Epithelioma	
1949	Nichelatti	F	16	4 months	Sarcoma	X-ray therapy, preceded by biopsy. No sign of recurrence two years later
1949	Barton	F	35	1 year	Basal-cell carcinoma	Excision and deep x rays. Later exenteration of orbit
1949	Toselli	M	55	2 years	Plasmoma	Excision

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Penman and Wolff grouped the cases according to the histological diagnosis and, although it is not very satisfactory to include in one table pathological opinions from different sources, the nine recent cases have been added to their list and regrouped into epithelial and non-epithelial tumours (Table II).

TABLE II
73 CASES PUBLISHED UP TO 1950

Epithelial				Non-epithelial			
Carcinoma	...	...	27	Fibroma	...	...	2
Papilloma	...	...	7	Sarcoma	...	...	22
Epithelioma	...	...	2	Endothelioma	...	...	4
Adenoma	...	...	1	Lymphoma	...	...	5
"Mixed" tumour	...	...	1	Plasmacytoma	...	...	2
Totals	...		38				35

Epithelial neoplasms, therefore, appear to be slightly more common than the non-epithelial and when it is realized that the diagnoses of "carcinoma", "papilloma", and "epithelioma" probably represent different stages of cylindrical-celled carcinoma, as we shall subsequently show, it will be seen that they form about 50 per cent. of all reported cases. The reasons for this opinion are based upon a clinical and pathological study of two recently encountered cases which represent primary tumours of epithelial origin but of differing degrees of malignancy.

FIG. 1.—Case 1. Drawing of the open sac showing a polypoid growth hanging from the upper pole.

CASE REPORTS

Case 1.—J. R., male, aged 46 years, van-driver, first noticed watering of the right eye. The tear duct had previously been syringed elsewhere, but he had felt no fluid pass through. After a month or two he became aware of a lump growing at the right inner canthus, and he came to Moorfields four months later.

Examination revealed a large cystic swelling closely resembling a mucocele of the lacrimal sac, but no mucus regurgitated on pressure. There was moderate epiphora. In July, 1949, a large thin-walled sac was removed. When the sac was opened a polypoid growth was found hanging from the upper pole (Fig. 1). The wound healed well but the lacrimal passages on this side are still not patent.

Pathological Findings.—Soft polypoid tumour measuring 32 x 15 x 11 mm.

Sections show a well-differentiated papillary tumour consisting of a richly-cellular, greatly-thickened cylindrical epithelium, in which numerous goblet cells and small retention cysts may be seen (Fig. 2). The epithelium, which consists of elongated fusiform cells (Fig. 3), proliferates inwards into the loose fibrous stroma in the centre of the tumour, and this ingrowth is particularly evident in the depths of the many irregular crypts (Fig. 2). The epithelium does not appear ciliated. The histological picture is that of a cylindrical-celled carcinoma of low-grade malignancy arising from the epithelial lining of the lacrimal sac.

FIG. 2.—Case 1. Cylindrical-celled carcinoma of low-grade malignancy. Papillary tumour consisting of greatly thickened cylindrical epithelium in which numerous goblet cells may be seen. Note that the epithelium proliferates inwards into the loose fibrous stroma in the centre of the tumour. Compare with Fig. 6. Masson stain x 39.

When the patient returned in January, 1951, no recurrence of the growth could be seen. He went on driving a van, and complained that his right eye was inclined to water only at the end of a long day of driving. It is perhaps worth emphasizing the minimal disability suffered by this patient although no communication exists between the conjunctival sac and the nose.

Case 2.—Y. S., female, aged 63 years, first attended Moorfields in July, 1949, complaining of a recurrent swelling at the inner angle of the left eye for the preceding

FIG. 3.—Case 1. High-power view showing the rich cellularity of the epithelium which consists of elongated fusiform cells. Compare Fig. 7. Masson stain $\times 122$.

few weeks. There was a yellowish discharge from the inner canthus, but very little watering and no pain.

Examination revealed a firm cystic swelling about 1 cm. square just below the inner canthus. Pressure on the swelling made a small amount of yellow fluid regurgitate from the lower punctum. On syringing, the left naso-lacrimal duct was not patent. A diagnosis of left mucocele was made and a dacryocystectomy carried out under local anaesthesia. Unfortunately the specimen was lost and its histological structure cannot be recorded.

The wound healed normally, but in January, 1950, the patient returned with a further swelling in the same site. The tumour was now discharging pus into the overlying skin, and epiphora had increased, but still there was no pain. As the discharge and swelling persisted, a diagnosis of infected residual sac tissue was made.

In August, 1950, she underwent a further left dacryocystectomy under general anaesthesia. Occupying the lacrimal fossa was a firm mass, indistinguishable from a lacrimal sac, and firmly adherent below and in front.

Pathological Findings.—Firm solid mass measuring 14 x 10 x 6 mm.

Sections show the tumour to consist of irregular masses of hyperchromatic cylindrical cells lying in a dense fibrous stroma lightly infiltrated with lymphocytes. Numerous goblet cells may be seen and in many areas these have distended to form cysts of varying sizes containing granular mucinous material. Within the larger cysts the epithelial cells have proliferated so as to impart a glandular appearance comparable with that of intra-duct carcinoma (Fig. 4). Mitoses are few but the growth is obviously malignant and it extends in all directions to the limits of the section. In its relatively solid areas the neoplasm is more squamous in type (Fig. 5). The histological picture is that of a highly malignant cylindrical-celled carcinoma of the lacrimal sac.

In view of these findings the patient's skull was x-rayed, with negative results, and she was referred to the ear, nose, and throat surgeon, Mr. G. H. Howells, who did not detect any neoplasm in the nasal or para-nasal cavities.

FIG. 4.—Case 2. Highly malignant cylindrical-celled carcinoma of lacrimal sac. Within cysts of varying sizes the epithelial cells have proliferated, imparting a glandular appearance comparable with that of intra-duct carcinoma. Masson stain x 84.

FIG. 5.—Case 2. Cylindrical-celled carcinoma. In its relatively solid areas the neoplasm is more squamous in type. Note the small cystic spaces which have developed from goblet cells. Compare with Fig. 8. x 160.

Fourteen days after operation there was already a fresh, firm swelling present beneath the lower part of the recent incision. In view of the patient's poor general condition it was considered that further surgery was to be avoided if possible and she was sent to the eye clinic at the Royal Cancer Hospital. In December, 1950, Dr. M. Lederman kindly supplied the following note:

The patient has had 5,600r in 47 days. The swelling shows considerable reduction but has not entirely disappeared. She is being kept under observation and if the growth shows signs of increasing, further surgical intervention may be undertaken.

DISCUSSION

Both from the clinical and pathological points of view these two cases form an interesting contrast. In the first case, an apparent cure followed a straightforward dacryocystectomy, and this benign outcome was in accord with the histological findings of low malignancy. In the second case, the neoplasm was much more malignant, and may have been provoked into further activity by incomplete removal.

Spratt (1940) has emphasized other writers' unanimous testimony that diagnosis is not possible in the first stage of the disease, because epiphora is the only symptom. In the second stage of swelling a diagnosis can be made. The presence of a round, hard mass, not reduced by pressure, especially if irrigation shows a patent duct, is indicative of thickening of the wall of the lacrimal sac rather than of a mucocele. In the third stage, when invasion of the lids and orbit has occurred, diagnosis becomes obvious. It is only in this stage that pain has been reported. The existence of Spratt's second stage may well be questioned, because it is unlikely that syringing would be successful in the presence of swelling and epiphora. Theoretically, one might have expected our first case, in which the polypoid tumour was suspended from the fundus of the sac, to have shown a patent duct on irrigation, for the growth could have caused epiphora and yet have been pushed aside by the lacrimal cannula, but actually syringing was never successful. Our second case should perhaps be classified somewhere between Spratt's second and third stages. The fact that the duct was not patent could be explained by the site of the tumour, near the outlet from the sac or from the top of the naso-lacrimal duct itself, or from an oedematous obstruction due to superadded infection.

From the pathological point of view it is of interest and importance to note that exactly similar tumours arise from the respiratory epithelium in the nasal and para-nasal cavities. This similarity, so far as we can ascertain, has not yet been sufficiently emphasized, but it is important, because the nasal tumours, although uncommon, are less rare than the corresponding growths in the lacrimal sac. Therefore attention to the natural history of these nasal tumours can perhaps help to explain the malignant potentialities of those arising

in the lacrimal sac, where their rarity is an obstacle to complete assessment.

This similarity had previously been noted by Muirhead (1933), who reported a case of lacrimal-sac carcinoma:

the section shows a transitional cell carcinoma, which is very similar in appearance to an *antral tumour*.

Verhoeff described the histological findings in a case reported by Spratt (1940), as follows:

. . . in a few places in the section there are remains of normal ciliated epithelium of mucous membrane. Such tumours may arise from *accessory sinuses*.

The illustrations of the histology in this article are exactly similar to those of our first case (Fig. 2). From an examination of two cases of polypoidal formation in the lacrimal sac, Tooke (1912) came to the conclusion that polyps of the tear sac, *as in the nose and accessory nasal cavities*, are evidence of hyperplastic growth or actual tumour formation which may possibly be due to a pre-existing inflammation.

NASAL TUMOURS.—These have been fully described by Kramer and Som (1935) and Ringertz (1938), and more recently by Lucas (1951). Kramer and Som collected a total of 81 cases from the literature, to which they added five of their own. They refer to the tumours as papillomata, a name criticized by Ringertz and Lucas; 70 per cent. of the cases were males and the tumours appeared at all ages but most commonly in the fourth decade. The experience of Ringertz (1938) was based on 111 cases of nasal papillomata, and his material showed that it is most convenient to divide them into two types, differing not only as regards their variety of epithelium but also in their architecture. The first type is composed of purely cylindrical epithelium, or of a transitional form between cylindrical and squamous, and is constructed quite differently from the ordinary papilloma of the skin. The second type has squamous epithelium and closely resembles the ordinary skin papilloma. It is the papillomata with cylindrical or transitional epithelium which correspond to the lacrimal-sac tumours. Ringertz reported 27 of these cases and pointed out that they should not be called papillomata, for the epithelium develops inwards, in contra-distinction to the true papilloma, in which it proliferates outwards. That is to say, they have an *inverted papillary structure* (Fig. 6). He called them “solid cylindrical carcinoma” and described three histological types:

- (1) typical cylindrical-cell carcinoma;
- (2) cylindrical-cell carcinoma with squamous metaplasia;
- (3) de-differentiated cylindrical-cell carcinoma.

Of the 27 cases reported, five showed local recurrences, metastasis, and local recurrence occurred in four cases, and metastasis only in four cases. The metastases occurred in both lymph glands and organs.

FIG. 6.—Solid cylindrical-celled carcinoma of nasal cavity. The epithelium is cylindrical in type and is proliferating inwards into the loose fibrous stroma. Compare Fig. 2. Haematoxylin and eosin. $\times 104$. (By courtesy of Dr. H. Lucas).

FIG. 7.—High-power view of Fig. 6. Note the cylindrical cells proliferating to form a greatly thickened epithelium. The picture is identical with that of the lacrimal-sac tumour. Compare Fig. 3. Haematoxylin and eosin $\times 147$. (By courtesy of Dr. H. Lucas).

In the series of 22 cases reported by Lucas (1951), the youngest was aged 22, and the oldest 77, with an average of 53 years. There were three females and nineteen males. The most common clinical manifestation was unilateral nasal obstruction with polypoid growth, which may be extensive and even protrude from the affected side. The growths usually arose from the nasal mucosa at a point between the ethmoid and antrum, whence they may extend into the ethmoid

cavity or the antrum or both. According to Lucas, the lesion begins as a widespread change in the respiratory epithelium, which undergoes metaplasia to a cylindrical type of cell. These cells are elongated, rather deeply staining and crowded together in palisade formation, resting on a basement membrane (Fig. 7). As the cells undergo de-differentiation they lose their cilia and their power to form mucous globules. This epithelial proliferation extends down into the ducts and mucous glands, and the growth is continued by an infolding of the epithelium into its own stroma (Fig. 6), so that solid masses of epithelium are gradually formed. This description conforms exactly with that of Ringertz. The cylindrical epithelium may then undergo squamous metaplasia and closely resemble a squamous-cell carcinoma (Fig. 8). The third type of growth, the de-differentiated cylindrical-cell carcinoma, did not occur in Lucas' series. Lucas agreed with Ringertz that the term papilloma should not be applied to these tumours, not only because their structure and mode of formation is different but also because such a name implies an innocent stage in their development, whereas they are probably malignant from the onset, varying only in the degree of their malignancy and corresponding clinical course.

TUMOURS OF THE LACRIMAL SAC.—It will already be clear from the above descriptions that we are here dealing with a tumour which is identical histologically and developmentally. Nor is this surprising, for it will be remembered that in the embryo the lining of the sac and the respiratory epithelium develop along parallel lines.

As Spratt (1937) pointed out, the normal lining of the lacrimal sac is composed of cylindrical cells. Repeated irritation, due to chronic inflammation, may cause this membrane to become thickened and to form several layers of cells which change to the squamous variety. Consequently sections may show the cylindrical, transitional, or squamous-cell type. The histology of our first case is exactly comparable with the early lesion in the nose, as a comparison of Fig. 2 with Fig. 6 and Fig. 3 with Fig. 7 will show. The second case is comparable with the "de-differentiated cylindrical-cell carcinoma" of Ringertz and shows also squamous metaplasia (compare Fig. 5 with Fig. 8).

It seems probable that all primary malignant epithelial tumours of the lacrimal sac arise in the same way, and that the different histological descriptions in the literature may be attributed to its varying developmental phases. Thus "carcinoma", "papilloma", and "epithelioma" were probably all examples of solid cylindrical carcinoma. In this connection the comments of Pasetti (1913) are of interest:

From what we were able to learn from these few observations of epithelioma of the lacrimal sac, all have been made up of cylindrical cells. All have exhibited marked malignancy. Recurrence is frequent.

FIG. 8.—Solid cylindrical-celled carcinoma of nasal cavity. The cylindrical epithelium has undergone squamous metaplasia. Compare Fig. 5. Haematoxylin and eosin x 138. (By courtesy of Dr. H. Lucas).

We have, therefore, seen by analogy with the nasal tumours that the primary epithelial growths of the lacrimal sac are probably all of the same nature, and that although they may 'appear benign histologically, they possess malignant potentialities of varying degree, and there is a strong tendency to recurrence. It is, therefore, clear that all nasal polyps and lacrimal tumours should be subjected to careful histological examination even if the clinical findings do not suggest malignancy.

SUMMARY

(1) Since the publication of the analysis of 64 reported cases of primary tumours of the lacrimal sac (Penman and Wolff, 1938), nine further cases have been found in the literature, and an analysis of the whole 73 cases shows that about 50 per cent. fall into the "papilloma-carcinoma" group.

(2) Two further cases of carcinoma of the lacrimal sac are reported.

(3) It is pointed out that the histology of these tumours is identical with that of solid cylindrical-cell carcinoma arising from the respiratory epithelium in the nasal and para-nasal cavities.

(4) The pathology of carcinomata of the nasal cavities and of the lacrimal sac have, therefore, been considered together, and the following conclusions reached:

(a) The term "papilloma" as applied to these growths should be abandoned, since their mode of formation and structure differs from that of a true papilloma.

(b) "Papillomata" arising from the lacrimal-sac epithelium are probably malignant from the beginning, as in the nose, and gradually or or rapidly progress to more obviously malignant cylindrical-cell carcinomata, a term which is, therefore, applicable to the whole group.

(5) The necessity for carrying out careful histological examination on all lacrimal tumours, even if the clinical findings do not suggest malignancy, is emphasized.

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