The second branchial arch

Rheumatological manifestations

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'And we have thought how the human fetus has, at one stage of its development, vestigial gill-slits' (JOHN STEINBECK: *The log from the sea of Cortez*).

In the course of normal development in man, the middle and largest part of the second branchial arch, the keratohyal cartilage, develops into a fibrous cord, the so-called stylohyoid ligament. Ossification takes place solely at its two ends, that is, in the hyoid bone and in the styloid process of the skull. In the present paper the significance for the clinical rheumatologist of some developmental anomalies of the styloid process will be considered.

Anatomy

The length of the styloid process is variable; the average length is usually assumed to be 2.5 cm. In this paper a styloid process longer than this will be referred to as 'long'.

COMPLETE OSSIFICATION of the branchial arch (Figs 1 and 2) is by no means rare; its occurrence has been known for more than 300 years and its history has been well reviewed by Franco, Aldrovando, and Goulart (1966). This is generally a harmless anomaly, though it may give rise especially in older subjects to a constrictive feeling in the throat or to phonasthenia. It may be mistaken for a swallowed chicken bone, and efforts to remove such a bone have been reported.

PARTIAL OSSIFICATION of the cranial part of the ligament (Fig. 3) will result in bony elongation of the styloid process, the site of fusion being often marked by a little knob, or, in some cases, by a radiotranslucent zone of connective tissue or cartilage.* If the length of the styloid process exceeds 3.5 cm., and especially if it is slightly bent inwards, pressure may

FIG. 1 Portion of skull, showing complete stylohyoid apparatus, with the hyoid bone somewhat upturned.

be brought to bear upon adjacent structures, notably blood vessels and nerves.

In seven random anatomical specimens, the distance from the base of the skull to the carotid artery measured: 3.9, 3.9, 3.8, 3.6, 2.2, 3.2, and 3.5 cm.

Apart from carotidynia (and even thrombosis of the internal carotid artery), hemicrania, neuralgia of the glossopharyngeal nerve or vagus, and sympathetic nerve symptoms may ensue. The rheumatologist who is consulted for indeterminate complaints in the neck, throat, or head must consider this frequently

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*Probably most cases of unilateral or bilateral (!) 'fracture' of the styloid process are instances of a partially ossified stylohyoid ligament.

FIG. 2 Radiograph, showing complete stylohyoid apparatus on one side and incomplete chain on the other.

overlooked possibility, which has been recorded in very few rheumatological papers (Layani, Fischgold, and Durupt, 1957; Layani, Durupt, and Nadaud, 1960; Wellinger, Dugast, and Desnos, 1966). Often, and for a considerable period, such patients are regarded as neurotics, until resection of the process per oram produces instant relief. In order to differentiate styloid pain from other conditions, an injection with novocaine in the tip of the process may be helpful.

A styloid process between 4 and 6 cm. long may reach the tonsil, and may then cause inconvenience before, during, and particularly after tonsillectomy. The rhinological literature deals with this problem.

The Finnish stomatologist Uotila (1965) reported an investigation of the length of the styloid process in 250 patients with rheumatoid arthritis and in 100 non-rheumatic controls. He found a long process to be present in 20 per cent. of the rheumatic subjects as compared with 10 per cent. of the controls. It should be mentioned, however, that these two groups were not matched for age and sex, a condition that must be fulfilled in view of the well-known predominance of rheumatoid arthritis in women and in persons over 40 years of age.

Incidence of elongated styloid process

In general, though the literature provides some information on the incidence of a long styloid process (see Discussion), data on the influence of age and sex are scanty. The present investigation has been carried out in two stages:

(1) Study of normal variations in the styloid process;

FIG. 3 Radiograph, showing partial cranial ossification of stylohyoid ligament; elongated styloid process. Knob-like locus of fusion.

(2) Measurement of the styloid process in rheumatic patients.

(1) Variations in the styloid process

The length of the styloid process was measured in 838 routine lateral radiographs of the cervical spine in out-patients of both sexes who were not suffering from rheumatoid arthritis and were more than 40 years old. Measurements were taken between the tip of the process and the point where it can be seen to emerge from under the base of the skull. A comparison between the measurements in anatomical specimens with those obtained from radiographs of the same skulls, revealed that in the radiographs the cranial part of the process remains hidden for a distance of 0.4 cm. On the other hand, using our technique for a lateral radiograph, the skull is enlarged 1.23 times, so that 0.48 cm. has to be added to the visible part of a process measuring 2.5 cm. of which only 2.1 cm. can be seen. The difference of 0.08 cm. was neglected. The size of the rest of the process, above 2.5 cm., has then to be divided by 1.23; as the same technique was used for all the radiographs, we refrained from doing this.

Surprisingly, a statistical analysis revealed that the incidence of a long process increased up to the age of 50 years in men and of 55 years in women, and then decreased in the higher age groups. There was no notable difference in the mean incidence between men and women (Table I overleaf).

Since all these patients had been x-rayed because of complaints in the shoulder girdle, these figures were compared with others obtained from x-ray

Table I *Incidence of styloid process more than 2.5 cm. long in persons over 40 years old*

Series	Total subjects	Age (yrs)	Non-rheumatic (838)				Rheumatic (123)			
			Male		Female		Male		Female	
			Total	Per cent. positive	Total	Per cent. positive	Total	Per cent. positive	Total	Per cent. positive
Out-patients	961	40-44	48	16.7	58	8.6	0	0	1	0.8
		45-49	47	23.4	79	13.9	5		5	60.0
		50-54	60	13.3	78	21.8	9	33.3	13	46.2
		55-59	57	10.5	73	15.1	15	33.3	20	60.0
		60-64	62	9.7	75	14.7	6	33.3	13	33.3
		65-69	44	6.8	54	9.3	2		17	64.7
		70+	44	5.4	59	8.4	5		12	50.0
		Total	362	12.1	476	13.7	42	26.2	81	54.3
Normal controls	175	40+		16.7		17.5				
Hospitalized rheumatoid patients	50	40+						33.3		39.4
Patients with ankylosing spondylitis	36	40+					24	40.4	12	44.4

films of the cervical spine in persons over 40 years of age in a study of a normal population (village of Bladel, Leiden Institute for Rheumatic Diseases). The incidence of a long styloid process in these healthy though possibly somewhat inbred persons proved to be slightly higher (Table I).

(2) *Measurement of the styloid process in rheumatic subjects*

Elongation of the process was found to occur more frequently in patients suffering from rheumatoid arthritis than in normal individuals, the mean incidence being approximately twice as high in men and four times as high in women. Moreover involution of the process after the age of 55 years failed to occur in women. Although it is probable that this also holds good for men, the number of male patients in this group was too small for valid statistical conclusions to be drawn (Table I).

The results of our observations in this series of 123 rheumatoid out-patients agree well with those obtained in a series of fifty patients who were more seriously affected and were hospitalized in the Leiden Department of Rheumatology, in whom the mean frequency was 33.3 per cent. in men, and 39.5 per cent. in women. The radiographs of these patients, and those in the population study, were studied and measured in collaboration with Dr. Cats of The Leiden Institute for Rheumatic Diseases (Table I).

In a small series of patients with ankylosing spondylitis who were more than 40 years old, the incidence was 40 per cent. in men and 44 per cent. in women (Table I).

In both rheumatic and non-rheumatic subjects the incidence of a long styloid process was inversely proportional to its size (Table II).

Table II *Incidence of increased elongation of styloid process in 961 out-patients over 40 years old*

Length of styloid process (cm.)	Controls (838)		Rheumatoid patients (123)	
	No.	Per cent.	No.	Per cent.
2.5+	106	12.6	54	43.9
4.0+	49	5.8	17	13.8
6.0+	13	1.5	15	12.2

Histology

The styloid process, with the stylohyoid ligament, of twelve patients who had died from non-rheumatic diseases was removed for histological examination. Metaplastic bone formation, both at the tip of the process and along its surface, was a constant finding (Figs 4 and 5, opposite). Part of the process frequently appeared as a hollow tube, and in some cases contained active bone marrow (Fig. 6, overleaf). Small fragments of cartilage, probably remnants of Reichert's branchial cartilage, were also encountered. These were usually completely embedded in bone, and small foci showing transition into bony tissue could sometimes be distinguished. Both decrease and increase in the length of the styloid process must therefore be considered to

FIG. 4 *Metaplastic bone along surface of styloid process. Haematoxylin and eosin. $\times 500$.*

FIG. 5 *Same specimen as Fig. 4, in birefringent light. Collagenous fibres can be seen to run into the bone without interruption. $\times 500$.*

FIG. 6 *Cartilage and cancellous bone around a lacuna, containing haematopoietic tissue. $\times 150$.*

depend on the metaplastic activity at its tip. In one specimen from a patient with rheumatoid arthritis (for which we are grateful to Prof. E. G. L. Bywaters) the picture was much more 'active'; the marrow cavity was large and the cortical bone layer was thin and in some places lacking, but no rheumatoid pathology could be seen.

Appearances in rheumatoid arthritis

No correlation was found between the length of the styloid process and the duration of rheumatoid arthritis or its activity, as assessed by the mean erythrocyte sedimentation rate during the last 3 years. It has already been pointed out that a long styloid process does not occur more frequently in severely affected patients. Nor could any qualitative or quantitative relationship be found with the Waaler-Rose test.*

Neighbouring joints, such as those of the cervical spine, were not more frequently affected than could be expected. The temporomandibular joint was involved in 35.2 per cent. in our patients with a long styloid process, and in 30.7 per cent. of those without.

*The Waaler-Rose test in ninety patients with a long styloid process, who did not suffer from rheumatoid arthritis, proved to be positive in 10 per cent.

There was a correlation of only 10 per cent. with ossification of the lesser horns of the hyoid bone, also reported to occur late in adult life.

We also studied the involution of the styloid process. The only other osseous process known to undergo considerable atrophy in old age is the alveolar process of the jaw; however, no correlation proved to exist between the length of the styloid process and atrophy of the alveolar process or the wearing of a dental prosthesis. Nor could an unusually high incidence of (clinical) deafness be demonstrated (the stapes bone is the most cranial part of the second branchial arch).

Discussion

As many authors fail to state what they mean by a 'long' styloid process, its incidence cannot easily be assessed from the literature, but the results are in fair accordance with each other, whether measured anatomically (von Eicken, 1919: 2 per cent.) or radiologically (Mangabeira-Albernaz, 1931: 3.3 per cent.; Eagle, 1958: 4 per cent.; Poggi Longostrevi and Milanesi, 1964: 2 per cent.; Hansen, 1965: 2.2 per cent.; Galmiche, Pallardy, Fournier, Bodson, and Pragier, 1966: a complete arch in 1.2 per cent.). Uotila (1965) used the term 'long process' for one reaching the hyoid bone. He used the planographic method, which will reveal thin bars of bone, not

detected on ordinary x-ray films; this also accounts for the high incidence recorded by this author.

Data in the literature relating the long styloid process to sex were hitherto almost nonexistent; our relevant data are listed in Table I. The incidence of a long process according to size is given in Table II. We found the elongation to be often symmetrical, and the preponderance of the left side, which was found by Hansen (1965), could not be demonstrated. Although our data were obtained by means of statistical analysis of information obtained at one point in time for each subject (transverse data), they have been confirmed in a limited number of longitudinal studies.

Our investigation indicates that the styloid process is more apt to change than might be supposed. Its greater length in elderly people had already been noted by early anatomists (Gruber, 1870; Sappey, 1876) whose findings have been more recently confirmed by Freese and Scheman (1962). The fact also appears in Uotila's tables, although the author does not comment upon it. On the other hand, Balestra and Chérie-Lignière (1903) in 200 anatomical specimens, Choppy (1955) in 100 radiographs, and Harburger (1925) in 500 subjects were unable to demonstrate this correlation. Our work confirms the older views.

As far as we have been able to ascertain, the opposite phenomenon (the subsequent involution of the process) has hitherto escaped attention.

Although Lucherini and Buratti (1967) described the frequent occurrence of frontal hyperostosis in patients with rheumatoid arthritis (also twice as frequent in women than in men), it would appear that, on the whole, patients suffering from rheumatoid arthritis do not exhibit a particular tendency towards ossification. It is true that patients with ankylosing spondylitis do show this tendency, but a long styloid process does not occur more frequently in them than in patients with rheumatoid arthritis; nor did we observe more than the expected number of long processes in our cases of vertebral hyperostosis or in our few patients with acromegaly.

With regard to the cause of the elongation of the styloid process, one is inclined to consider the possibility of a congenital atavistic anomaly, but this hypothesis cannot easily be proved. In children, up to adolescence, the styloid process, being still cartilaginous, is invisible radiographically. We have, however, examined three generations of a large family, and we have found indications for an autosomal dominant mode of inheritance. Hormonal and inflammatory influences have been held responsible, as well as mental debility and psychopathy (Chérie-Lignière, 1906), brachycephaly (Cimino, 1955), race (Eagle, 1958), and strain on the vocal cords (Corsy, 1920).

Although the association of a long styloid process and rheumatoid arthritis is undeniable, we are unable to offer an explanation for this relationship. As elongation of the styloid process, apart from its mechanical effects, is symptomless, there is as yet insufficient reason for coining the clinical term 'second arch syndrome', analogous to the 'first arch syndrome' (François and Haustrate, 1954; McKenzie, 1966).

Summary

Partial or complete ossification of the stylohyoid ligament, a remnant of the second branchial arch, is not rare. If partial ossification results in elongation of the styloid process, fairly characteristic symptoms may ensue, through pressure upon blood vessels or nerves.

Normal variations in the length of the styloid process, in relation to age and sex, have been ascertained by the investigation of more than 1,000 normal subjects. The incidence of a long process shows a steady increase with advancing age until 45 to 50 years in men, and 50 to 55 years in women, with a subsequent decrease in older subjects.

Histological examination revealed metaplastic bone formation in the adult process.

In a series of 123 patients with rheumatoid arthritis, on the other hand, the incidence of a long process was found to be twice as high in men and four times as high in women than in normal controls. No statistical correlation between the length of the process and various aspects of rheumatoid arthritis were found. The only analogy in the literature is the high incidence of frontal hyperostosis in patients with rheumatoid arthritis.

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Discussion

PROF. E. G. L. BYWATERS (*Taplow*) Any involutionary process in bone probably depends on collagenase. Is it possible that there was some correlation with treatment—certain treatments being known to affect collagenase activity? Was there any relationship with the type or duration of treatment, aspirin usage, steroids, chloroquine, gold, or the absence of any particular form of treatment?

DR. DE HAAS We could not find any correlation with any treatment used.

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RÉSUMÉ

La seconde arche branchiale Manifestations rhumatismales

L'ossification partielle ou complète du ligament stylohyoïdien, un vestige de la seconde arche branchiale, n'est pas rare. Si l'allongement de l'apophyse styloïde résulte de l'ossification partielle, des symptômes plutôt caractéristiques dus à la pression sur les vaisseaux sanguins et les nerfs peuvent s'en suivre.

Les variations normales de la longueur de l'apophyse styloïde en relation de l'âge et du sexe ont été obtenues par l'examen de plus de 1.000 sujets normaux. L'incidence d'une longue apophyse montre une augmentation graduelle avec l'âge jusqu'à 45 à 50 ans chez l'homme, et 50 à 55 ans chez la femme, et une diminution subséquente chez les sujets plus vieux.

L'examen histologique a révélé la formation d'os métaplastique dans l'apophyse des adultes.

Par contre, dans une série de 123 malades atteints d'arthrite rhumatoïde, l'incidence d'un long apophyse a été trouvée comme étant deux fois plus élevée chez les hommes et quatre fois plus élevée chez les femmes que chez les témoins normaux. Aucune corrélation statistique entre la longueur de l'apophyse et les divers aspects de l'arthrite rhumatoïde n'a été trouvée. La seule analogie dans la bibliographie est la haute incidence de l'hyperostose frontale chez les malades atteints d'arthrite rhumatoïde.

SUMARIO

El segundo arco branquial Manifestaciones reumatológicas

La osificación parcial o total del ligamento estilohioideo, un vestigio del segundo arco branquial, no es rara. Si la osificación parcial produce elongación del proceso estiloide, podrían seguir síntomas bastante característicos, debido a presión sobre los vasos sanguíneos o nervios.

Se han determinado las variaciones normales en la duración del proceso estiloide, con relación a edad y sexo, mediante el estudio de más de mil sujetos normales. La incidencia de un proceso largo revela un aumento continuo con el paso de los años hasta los 45 a 50 años en los hombres, y 50 a 55 en las mujeres, con una disminución subsiguiente en sujetos mayores.

El examen histológico reveló formación de hueso metaplastico en el proceso adulto.

Por otro lado, en una serie de 123 pacientes con poliartritis reumatoide, se descubrió que la incidencia de un proceso largo era dos veces mayor en los hombres y cuatro veces más alta en las mujeres, comparado con testigos normales. No se halló ninguna correlación estadística entre el tamaño del proceso y los varios aspectos de la poliartritis reumatoide. La única analogía en la literatura es la alta incidencia de hiperostosis frontal en pacientes con poliartritis reumatoide.