Original Article

Pattern of Ear Diseases in Surgical Pathology

Ear Diseases in Surgical Pathology

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ABSTRACT

Objective: The purpose of this study is, firstly, to find the pattern of ear diseases in the community, as no such pattern study is currently available. Secondly, to impart awareness regarding microscopic appearances of the common ear diseases encountered in this pattern study.
Study Design: Observational / descriptive study.
Place and Duration of Study: This study was conducted at the Surgical Pathology archives of the Laboratory of Charsada Teaching Hospital affiliated with Jinnah Medical College Peshawar from 2010 to 2015.
Materials and Methods: In this study, all the cases of ear diseases from surgical pathology archives of the laboratory of Charsada Teaching Hospital were retrieved. All the slides and the diagnoses for the retrieved cases were reviewed by the histopathologist and the final diagnoses were recorded; the disease pattern was determined, keeping in view the number of cases for each diagnostic category and the patient's age.
Results: The review of these cases between 2010 and 2015 showed that most of the biopsy specimens were from external ear while few were from the middle ear. The first five commonest conditions constituted almost two thirds of the total number of cases. The congenital anomalies and juvenile xanthogranulomas were most common in the first decade of life. Benign and malignant tumors were uncommon and seen mostly after the third decade of life.
Conclusions: Congenital anomalies and inflammation associated diseases are common in the first three decades of life, whereas benign and malignant neoplasms are more common after the third decade of life.
Key Words: Accessory tragus, cholesteatoma, keratosis obturans, angiolymphoid hyperplasia with eosinophilia, ear diseases.

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INTRODUCTION

Ear diseases are quite common in clinical practice; however, ear biopsies are not frequently encountered in surgical pathology practice. External ear and to some extent middle ear are accessible to biopsy procedure. External and middle ear are composed of bone and cartilage covered over by muscles, soft tissue and skin with adnexa. Therefore, ear is prone to all the diseases that can affect these components structures individually. Usually, masses or non-resolving inflammatory lesions will be biopsied to ascertain their nature. The purpose of this study is, firstly, to find the pattern of ear diseases in the community, as no such pattern study is currently available. Secondly, to impart awareness regarding microscopic appearances of the ear diseases encountered in this pattern study.

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MATERIALS AND METHODS

In this laboratory based retrospective study, all the cases of ear diseases between the year 2010 and 2015 from surgical pathology archives of Charsadda Teaching Hospital affiliated with Jinnah Medical College Peshawar were retrieved. All the ear biopsy cases were considered irrespective of patient age, gender, or ethnic origin. As a first step all the slides and the diagnoses for the retrieved cases were reviewed and the final diagnoses were noted down; the second step involved recording the number of cases for each diagnostic category; and as a final step, all the diagnoses were tabulated in order of most common to least common, and the disease prevalence tabulated according to the patient's age.

RESULTS

The review of cases of ear disease between 2010 and 2015 showed that most of the biopsy specimens were from external ear while few were from the middle ear, Table 1. Congenital anomalies and juvenile xanthogranulomas were most common in the first decade of life. Inflammatory aural polyp was common in the second decade. Cholesteatoma straddled the second and third decades; while keratosis obturans was straddling the third and fifth decades. Angiolymphoid hyperplasia with eosinophilia was common in the third decade.

Benign adnexal tumors were common in the fourth and fifth decades while malignant tumor (squamous cell carcinoma) was seen in the ninth decade.

Table No.1: %age of ENT diseases.

Sr. No.	Diseases	Number of cases (n)	%age
1.	Congenital anomalies	23	22.5
2.	Epidermoid cyst	20	19.6
3.	Cholesteatoma	15	14.7
4.	Keratosis obturans	10	9.8
5.	Inflammatory aural polyp	9	8.8
6.	Juvenile Xanthogranuloma	7	6.9
7.	Keloid	4	3.9
8.	Angilymphoid hyperplasia with eosinophilia	3	2.9
9.	Seborrheic keratosis	3	2.9
10.	Chondrodermatitisnodularis	2	1.9
11.	Nodular fasciitis	2	1.9
12.	Fungal infection	1	0.98
13.	Trichoepithelioma	1	0.98
14.	Syringoma	1	0.98
15.	Squamous cell carcinoma	1	0.98
	Total Cases	102	

DISCUSSION

Surgical specimens from the external and middle ear are fairly uncommon in hospital based surgical pathology practice. Main indications for biopsy from this site being: mass lesion, an eroding lesion, or non-resolving inflammation.

In our study, most of the cases belonged to the congenital anomaly category, encompassing mainly accessory tragi and pre-auricular sinuses (n=23, 22.5%), Table 1. These were most common in the first decade of life Table 2, as the alarmed parents brought their children (with obvious abnormalities near the ear) for early consultation. Microscopically accessory tragi were composed of fibroadipose tissue with or without central cartilage and covered by skin with adnexal structures. The preauricular sinuses were composed of stratified squamous epithelium-lined sinus tract surrounded by lymphocytes and plasma cells. Both are anomalous derivatives of first and second branchial arches appearing as skin colored nodules (sessile or pedunculated) or pits (preauricular sinuses) in front of the ears. Though accessory tragi are benign, they may be associated with certain syndromes like Delleman's syndrome, Goldenhar's syndrome, Haberland's syndrome, and Townes-Brock's syndrome[1]. Therefore, it is advisable to assess the neonate/infant with branchial arch anomalies for any associated renal, ocular, vertebral, and brain anomalies.

The second most common ear biopsy diagnosis in the study was epidermoid cysts (n= 20, 19.6%), Table 1. The commonly affected sites were the pinna and outer external auditory canal. It presented as a nodule at the site of trauma or ear piercing. They were common in third and fourth decades of life Table 2. They were characterized by a cyst wall lined by keratinized stratified squamous epithelium replete with granular cell layer, the cyst lumen contained keratin flakes. Epidermoid cysts though totally benign, may be part of Gardner's syndrome especially if multiple cysts are found all over the body[2,3,4,5]. Therefore when multiple epidermoid cysts are encountered all over the body, colonoscopy to detect the presence of colonic polyps is mandatory.

Cholesteatoma (n= 15, 14.7%), the third common biopsy diagnosis, mostly occurred in the middle ear (11 cases) with only two cases in the mastoid and two cases in the external auditory canal. Chronic ear discharge and conductive deafness were the main complaints. Microscopically it was characterized by multiple keratin flakes mostly admixed with acute inflammatory cells, with thin to normal thickness keratinizing stratified squamous epithelium with variable amount of acute and chronic inflammatory cells in the sub-epithelial stroma. It was common in the second and third decades of life Table 2, though literature states it to be common in the first and second decades. This depicts difference in the age groups affected by preceding otitis media in different communities. Its importance lies in the fact that in spite of being benign, it is an erosive and an expansive lesion that may result in bone necrosis, cavitation and remodeling; therefore, it merits regular follow-up[2,4,6].

Keratosis obturans (n= 10, 9.8%), the fourth common biopsy diagnosis was common in the third and fifth decades of life Table 2. It mostly presented with conductive deafness Table 1. It was composed of compact keratin flakes; the lining epithelium of affected external auditory canal was variably acanthotic showing hyperkeratosis and parakeratosis. Contrary to cholesteatoma, this is an innocuous, non-eroding and non-cavitating lesion. It is said to results from failure of the self-cleaning mechanism of the external auditory canal due to some unknown cause[4]. Evacuation of the keratinous ball is all that is required for the management.

Inflammatory aural polyp (n=9, 8.8%) was the fifth common biopsy diagnosis, mostly affecting the middle ear and presented as a soft, easy to bleed, polypoid granulation tissue associated with middle ear infections; therefore, seen commonly in the second decade Table 2. Microscopically, it was usually partially covered by ulcerated cuboidal to stratified squamous epithelium with underlying lobular capillary proliferation, the stroma being heavily infiltrated by acute and chronic inflammatory cells[4]; the ulcerated portion was covered by acute inflammatory exudate with few superficial bacterial colonies.

Table No.2: ENT diseases – decade wise

Diseases	Decades of Life									
	1st	2nd	3rd	4th	5th	6th	7th	8th	9th	Total
Congenital anomalies	14	4	3	2	-	-	-	-	-	23
Epidermoid cyst	2	3	4	7	3	-	1	-	-	20
Cholesteatoma	1	6	4	2	1	1	-	-	-	15
Keratosis obturans	-	1	3	1	5	-	-	-	-	10
Inflammatory aural polyp	1	4	2	-	1	-	-	1	-	9
Juvenile Xanthogranuloma	5	2	-	-	-	-	-	-	-	7
Keloid	-	1	3	-	-	-	-	-	-	4
Angilymphoid hyperplasia with eosinophilia	-	-	2	-	1	-	-	-	-	3
Seborrheic keratosis	-	-	-	-	-	2	1	-	-	3
Chondrodermatitisnodularis	-	-	-	-	-	-	-	2	-	2
Nodular fasciitis	-	-	1	1	-	-	-	-	-	2
Fungal infection	-	-	-	-	-	1	-	-	-	1
Trichoepithelioma	-	-	-	-	1	-	-	-	-	1
Syringoma	-	-	-	1	-	-	-	-	-	1
Squamous cell carcinoma	-	-	-	-	-	-	-	-	1	1

Juvenile xanthogranuloma (n= 7, 6.9 %) was the sixth common biopsy diagnosis received. It was found to be common in the first two decades Table 2, presenting as a pink- to red- to tan nodules on the skin. Microscopically, it was composed of circumscribed infiltration by foamy macrophages, multinucleate Touton type of giant cells, lymphocytes, and scattered few eosinophils. Two thirds of these cases are reported to occur in the first year of life, mostly presenting as a solitary lesion[7].

Keloid (n= 4, 3.9%) was the seventh common biopsy diagnosis in this study, more common in the third decade, occurring at the ear lobule and pinna, mostly secondary to trauma or ear piercing. The dermis showed many irregularly arranged thick hyalinized collagen bundles in hypocellular matrix without adnexal structures. The epidermis was mostly atrophic. Spontaneous occurrence of keloids has been reported in diseases like Ehlers-Danlos syndrome type IV, Rubenstein-Taybi and Gorlin syndrome, and scleroderma[4].

The eighth common biopsy diagnosis was shared by angiolymphoid hyperplasia with eosinophilia and seborrheic keratosis, each represented by 3 cases, Table 1. Angiolymphoid hyperplasia with eosinophilia (n= 3, 2.9%) Table 1, also known as epithelioidhemangioma, was found to affect the skin of pinna, immediate periauricular area, and outer part of external auditory canal. It presented clinically as pruritic cutaneous nodules in third decade of life Table 2. Microscopically, it showed dermal lymphoid hyperplasia marked by lymphoid aggregates, lymphoid follicles, and interstitial lymphoplasmacytic infiltrate admixed with macrophages and eosinophils. There were many scattered thick-walled blood vessels lined by prominent epithelioid endothelial cells, containing enlarged hyperchromatic nuclei. The epidermis was mostly unremarkable. This disease needs to be differentiated from Kimura's disease which is characterized by subcutaneous location accompanied by regional lymphadenopathy, blood eosinophilia, and increased IgE levels[2,3,4,5]. Seborrheic keratosis (n=3, 2.9%) was common in the sixth decade Table 2. It presented as greasy exophytic lesions located on the pinna. Microscopically, there was regular epidermal acanthosis due to proliferation of basaloid cells with many true and pseudohorn cysts. It should not be confused with basal cell carcinoma, which is characterized by irregular proliferation of basaloid cells extending into the dermis with surrounding clefts and degenerative dermal changes. Those lesion, which suddenly increases in size or number, may be associated with an internal malignancy (Leser-Trelat sign)[7].

The ninth common biopsy diagnoses were chondrodermatitisnodularishelicis and nodular fasciitis, each represented by two cases, Table 1. Chondroder-matitisnodularishelicis (n= 2, 1.9%) was seen in the eighth decade Table 2, presenting as an ulcerated nodule over the helix of pinna. Microscopically, it showed central ulceration covered by inflamed granulation tissue extending down to involve the cartilage. The surrounding intact epidermis showed acanthosis, hyperkeratosis, and parakeratosis. There were focal upper dermal changes of solar elastosis. It is a disease of unknown etiology; the suggested etiologies include ischemia, pressure, trauma, actinic, cold, or immune mediated injury[4,7]. Nodular fasciitis (n= 2, 1.9%) was seen affecting the pinna and outer part of the auditory canal in the third and fourth decades Table 2, it presented as an enlarging mass of two to three weeks duration. Microscopically, it was occupying the dermis and subcutaneous tissue, composed of loosely arranged spindle shaped cells in variable myxoidstroma with undulating collagen bundles with some infiltration by mature lymphocytes.

The tenth position was shared by four biopsy diagnoses, namely fungal infections, trichoepithelioma, syringoma and squamous cell carcinoma; these diagnostic categories were represented by one case each. Fungal

infection (n=1, 0.98%) presented in sixth decade in an old diabetic patient Table 2, characterized by poor disease control accompanied by intractable headache and ear discharge. The middle ear biopsy showed necrotic bone fragments with scattered clusters of broad ribbon-like hyphae with parallel but undulating walls and irregular branching. Also, there was acute and chronic inflammatory infiltrate with scattered multinucleate giant cells with some ingested hyphae. In ambiguous situations staining the sections with PAS (Periodic Acid Schiff) and GMS (Grocott's Methenamine Silver) stains will highlight the presence of fungi[8].Trichoepithelioma (n=1, 0.98%) presented as a solitary, skin colored, cutaneous nodule over the ear lobule in the fifth decade Table 2. Microscopically, there was a circumscribed dermal proliferation of basaloid cells with peripheral cellular palisading and small petal-like protrusions, surrounded by some fibromyxoidstroma, without formation of hair. Multiple trichoepitheliomas may occur as an autosomal dominant familial condition or be part of Brooke–Spiegler syndrome when together with multiple cylindromas[7]. Syringoma (n= 1, 0.98%) was seen as a small pale skin nodule at the tragus, in the fourth decade, Table 2. Microscopically, it showed upper dermal proliferation of tubular structures of different sizes, some with comma-like solid cellular extensions. The tubular structures were lined by double layer of cuboidal cells, with some luminal secretions. It should be remembered that multiple syringomas may be seen in Costello syndrome and Down's syndrome[4]. Squamous cell carcinoma (n=1, 0.98%) presented as an ulcerated 1 cm nodule over the upper part of right helix in the ninth decade Table 2. Microscopically, it showed pleomorphic malignant squamous cells invading the upper dermis with formation of keratin pearls, but without involvement of the perichondrium, cartilage or the regional lymph nodes. The adjacent dermis showed solar elastosis and mild lymphoplasmacytic infiltration. The chance of involvement of the regional lymph nodes has been found to be directly proportional to the primary tumor size; moreover the disease prognosis depends on the tumor stage[2].

There is no available prior national or international study for pattern of ear diseases in surgical pathology. However, some clinical studies for ear diseases are available. In a ten year hospital-based retrospective study for geriatric patients, conducted at the ENT Department of University College Hospital, Ibadan, Nigeria[9], it was found that impacted cerumen, hearing loss, and chronic suppurative otitis media were the three most common diseases of the ear in elderly. In another prospective cross-sectional study among 2000 children (aged between 5 and 13 years), it was found that impacted wax followed by chronic suppurative otitis media and otitis media with effusion were the most common ear diseases in rural school children of Nepal[10]. A similar study, conducted in Kalyanpuri, Delhi, a total of 1398 school children (aged between 5 and 14 years) were studied for ear diseases, impacted wax was the commonest problem (23.4%) followed by ear infections (10.0%) and hearing impairment (7.2%)[11].

CONCLUSION

The congenital anomalies along with inflammation associated diseases are most common in the first three decades of life; being responsible for two thirds of all the surgical pathology biopsies of the ear, whereas, neoplasms are more common after the third decade of life.

Conflict of Interest: The study has no conflict of interest to declare by any author.

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