Profile of Schwannoma in a Tertiary Orthopedic Center- Seven Year Experience

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Abstract:

Context: Schwannomas are rare benign soft tissue tumours. It presents as painless, slowly growing swelling. It may involve any nerve in the body. There are few published data and literature on Schwannoma. Hence, we took up this study to analyze the pattern of Schwannoma.

Aims: To study the clinico pathological features of Schwannomas.

Settings and Design: Retrospective analysis of Schwannoma diagnosed from February 2015 to February2022 at Sanjay Gandhi Institute of Trauma & Orthopedics, Bangalore.

Methods and Material: The present study comprised of retrospective analysis of Schwannoma diagnosed from February 2015 to February2022 at Sanjay Gandhi Institute of Trauma & Orthopedics, Bangalore. Total of 22 biopsies were analyzed. Relevant clinical and radiological details were obtained. Biopsy specimens were processed routinely and wherever necessary special stains and IHC for S-100 were done.

Statistical analysis used: Nil

Results: In our study, the common age groups affected were between 41-50years. Men were affected more. Most common symptoms were pain and swelling. Youngest was 17 year old male and oldest was 72 year male

Conclusions: Understanding the morphological variants of schwannomas aids the pathologist in making a precise diagnosis

Keywords: Histopathology, Schwannoma.

Key Messages: Schwannomas are rare benign neoplasm that typically appears as a single, solitary asymptomatic lump anywhere on the body. Schwannoma should be considered in the differential diagnosis of any slow growing asymptomatic lesion. It has a wide range of morphological variation and offers a challenge in the diagnosis. Understanding the morphological variants of schwannomas aids the pathologist in making a precise diagnosis.

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Introduction:

Schwannomas are rare benign soft tissue tumours. The synonymous terms are neuroma or neurilemmoma. Verocay had labelled the tumour as neurinoma in 1910. Masson introduced the term "Schwannoma" in 1932.Later, in 1935, the term neurilemmoma was coined by Stout. [1] Any cranial, peripheral or autonomic nerve containing Schwann cells, the sheath cells that encase the myelinated nerve fibers, is susceptible to the development of schwannomas[2].The classical schwannomas are well circumscribed, encapsulated masses. Often the associated nerve can be identified without invaded by Schwannoma. Grossly, they appear as firm, gray masses. Frequently, a related nerve can be seen stretched over its peripheral side. Cut sections reveal, firm, light, glistening tissue, with focal areas of haemorrhage, cyst formation. Microscopically, biphasic pattern, that is cellular Antoni A areas alternating with hypocellularAntoni B areas and distinctive Verocay bodies are seen. [3] They often show up , especially in long standing cases, degenerative changes that can be very pronounced. These include nuclear atypia (without increased mitotic activity), prominent hyalinised blood vessels, foamy histiocytes and others[4] .

Schwannomas affect all ages and are most common in fourth to sixth decade of life. They do not show any predilection for race or sex. Over 90% of these lesions are single and sporadic. Most common sites include peripheral nerves in skin and subcutaneous tissue of head and neck, along the flexor surface of extremities, spinal nerve roots forming Intradural, extramedullary tumours. Rare locations are spinal intra medullary, CNS, GIT and bone [5].

WHO has classified schwannomas as conventional, cellular, plexiform, micro cystic/reticular, ancient subtypes [5]. Various subtypes in schwannomas pose difficulties in histopathological diagnosis [4, 5 and 6]. Our study aims to determine the clinico pathological characteristics, subtypes of Schwannoma seen in tertiary orthopaedic hospital.

Subjects and Methods: A retrospective, cross sectional, hospital based study was conducted at Department of Pathology, Sanjay Gandhi Institute of Trauma &Orthopedics, Bangalore, Karnataka, India. The study consisted of 22 cases of schwannomas diagnosed in the Histopathology department during period of seven years (Feb 2015-feb 2022). The biopsy samples were fixed in 10% buffered neutral formaldehyde. Tissue was processed by increasing concentrations of alcohol and paraffin blocks were prepared. Sections were cut to 4-6µ, stained by haematoxylin and eosin and examined under microscope for histopathological examination. Special stains and immunohistochemical studies were performed wherever necessary. The final diagnosis was made. The clinical and pathological data were collected from medical records and reviewed for patient demographics, age, and sex and radiophraphic details. Data tabulation and analysis done to know the relative frequencies of observed data. Results are expressed as numbers and percentages.

Results:

In the study period of seven years from February 2015 to February 2022, 22 cases of schwannomas were diagnosed.

Table 1 shows the distribution of Schwannoma by age and gender. Schwannomas in present study, ranged from first to 8th decade. The common age group affected was between 41-50 years. Youngest patient was 17 years old and oldest was 72.

years. It is more common in men.

Age group	Male	Female	number
10-20	02	00	02
21-30	05	02	08
31-40	02	00	02

41-50	05	04	09
51-60	02	00	02
61-70	00	00	00
71-80	01	00	01
Total	16	06	22

Table 1: Age and Gender distribution of Schwannoma

Sl no.	Location	Number of cases
01	Intradural extra medullary- cervical Thoracic Lumbar	02 02 05
02	Extra Dural mass -L_5-S_1	02
03	Antrochoanal area	01
04	Toe	01
05	Forehead	02
06	Left ring finger	01
07	Sural nerve	01
08	Gluteal region	01
09	Site not specified	04
Total		22

Table 2: Locations of Schwannoma

Table 2: describes various locations of Schwannoma.

In the present study, 9 (40.90%) of the cases were located in spine. Majority of the these were intramedullary and located in lumbar area. Site was not mentioned in 4(18.18%) cases. We could not get the details of these patients as these were referred cases.

		Features							
	Type	Capsule	Antoni A	Antoni B	Verocay bodies	Cystic areas	Hyalinised blood vessels	Inflammatory cells	Atypical cells
1	Cellular	+	-	+	-	-	-	-	+
2	Cellular	+	+	+	-	-	+	+	-
3	Cellular	+	+	+	+	-	+	-	+
4	Cellular	+	+	+	-	-	+	-	+
5	Cellular	+	+	+	-	-	-	-	-
6	Cellular	-	+	+	-	-	+	-	-
7	Cellular	-	+	+	-	+	+	+	+
8	Cellular	-	+	+	-	-	+	-	-
9	Classical	+	+	+	+	-	+	+	-

10	Classical	+	+	+	+	-	+	-	-
11	Classical	-	+	+	+	-	-	-	-
12	Classical	+	+	+	+	-	+	-	-
13	Classical	-	+	+	+	-	+	+	-
14	Classical	+	+	+	+	-	+	+	-
15	Classical	+	+	+	+	-	+	+	-
16	Plexiform	+	+	+	+	-	+	+	-
17	Plexiform	+	+	+	+	-	-	-	-
18	Ancient	+	+	+	-	-	+	+	+
19	Ancient	+	+	+	-	-	+	+	+
20	Pseudoglandular pattern	+	+	+	-	+	+	-	-
21	Pseudoglandular pattern	+	+	+	-	+	+	+	-
22	Pseudoglandular pattern	+	+	+	-	+	+	Haemosiderinlayden macrophages	-

Table : 3 – Histopathological features of Schwannoma

Table 3 describes the histopathological features of schwannoma. Microscopically the characteristic features of Schwannomas were seen in all the cases. 18/22, (81.81%) of cases were encapsulated. Mixture of AntoniA&Antoni B areas were found in all the cases. Microcyst formation was observed in 22.72 %.

Verocay bodies were seen in 50% of the cases.Degenerative changes composed of nuclear atypia, pleomorphism was observed in 27.27 %.

Sl no	Variant	No of cases	Percentage of cases
1	Cellular	08	36.36%
2	Classical	07	31.81%
3	Pseudo glandular pattern	03	13.63%
4	Ancient	02	9.09%
5	Plexiform	02	9.09%
6	Total	22	100%

Table : 4 - Variants of Schwannoma

Cellular schwannomas constituted major type contributing around 36% of the cases, plexiform and ancient pattern constituted 9.09% of cases each.

Sl no	Schwannoma	Age/Sex	Site	Clinical diagnosis
1	Cellular	23/M	Gluteal region	Neuro fibroma
2	Cellular	43/M	D_9 Lesion	Schwannoma
3	Cellular	27/F	L_5 S_1 Lesion	Schwannoma
4	Cellular	23/M	$L_3 – L_4$	Meningioma
5	Cellular	72/M	Sural nerve	Not known
6	Cellular	44/M	Antrochoanal polyp	Nasal polyp
7	Cellular	25/M	Not known	Not known
8	Cellular	33/M	Forehead	Not known
9	Classical	49/F	Not known	Neurofibroma
10	Classical	42/F	Lumbar	Neurofibroma
11	Classical	20/M	Forehead	Neurofibroma
12	Classical	55/M	Lumbar	Neoplasm of unknown behaviour
13	Classical	21/M	$L_3 –L_4$	Schwannoma

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14	Classical	27/M	Cervical spine	Schwannoma
15	Classical	50/M	Toe	Not known
16	Pseudoglandular pattern	52/M	$L_1 – L_2$	Neuro fibroma
17	Pseudoglandular pattern	43/F	Not known	Neuro fibroma
18	Pseudoglandular pattern	48/F	L_1	Neuro fibroma
19	Ancient	50/M	D_{10}- D_{11} lytic lesion	Mass lesion compressing the cord
20	Ancient	30/F	D_{11} - D_{12} lytic lesion	Schwannoma
21	Plexiform	17/M	Left ring finger	Not known
22	Plexiform	36/M	C_1	Schwannoma

Table: 5 Clinical features of Schwannoma.

Table 5 describes the Clinical diagnosis rendered in the cases diagnosed as schwannoma histopathologically.Clinical diagnosis was available in 18/22 cases. Most common clinical diagnosis of the lesions was Neurofibroma in 7/18 cases. Only in 6/18 cases clinical diagnosis correlated with histopathological diagnosis

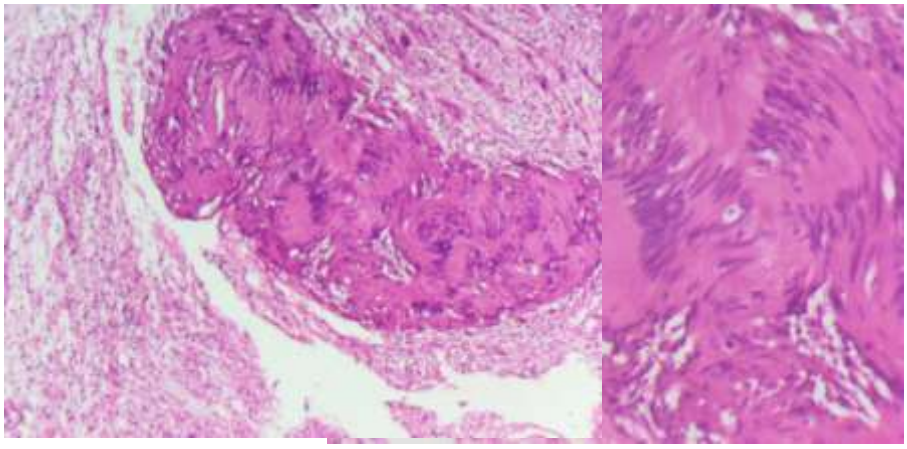

Figure 1: Spindle to elongated cells arranged in whorls with AntoniA Antoni B pattern (H and E, x 04) along with Verocay bodies (H and E, x 400).

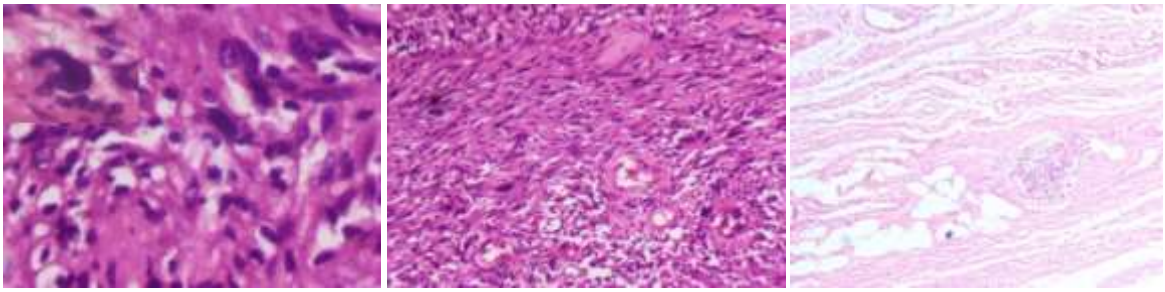

Figure 2: Cell pleomorphism (H&E, x 400), hyalinised blood vessels (H&E x 100)Intracapsular collections of lymphocytes, (H&E, x 40) in ancient schwannoma.

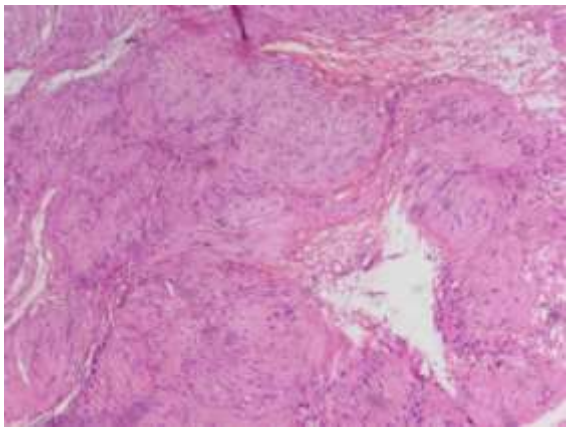

Figure 3: Spindle to elongated cells arranged in bundles and in clusters separated by thin fibrous bands in plexiformschwannoma(H & E x 40)

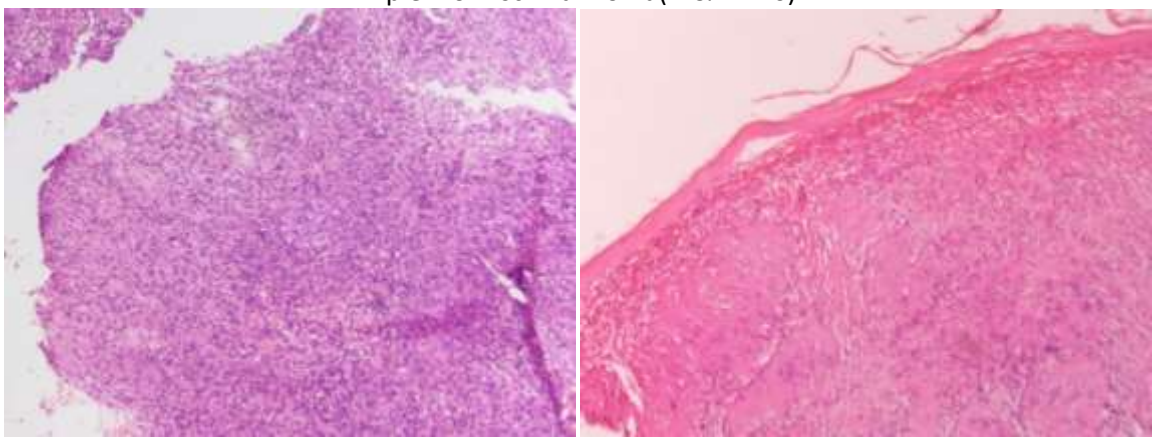

Figure 4: Schwannoma of the nasal cavity lacking capsulation. (H & E, x40), plexiformschwannoma showing capsule. (H&E, X 40)

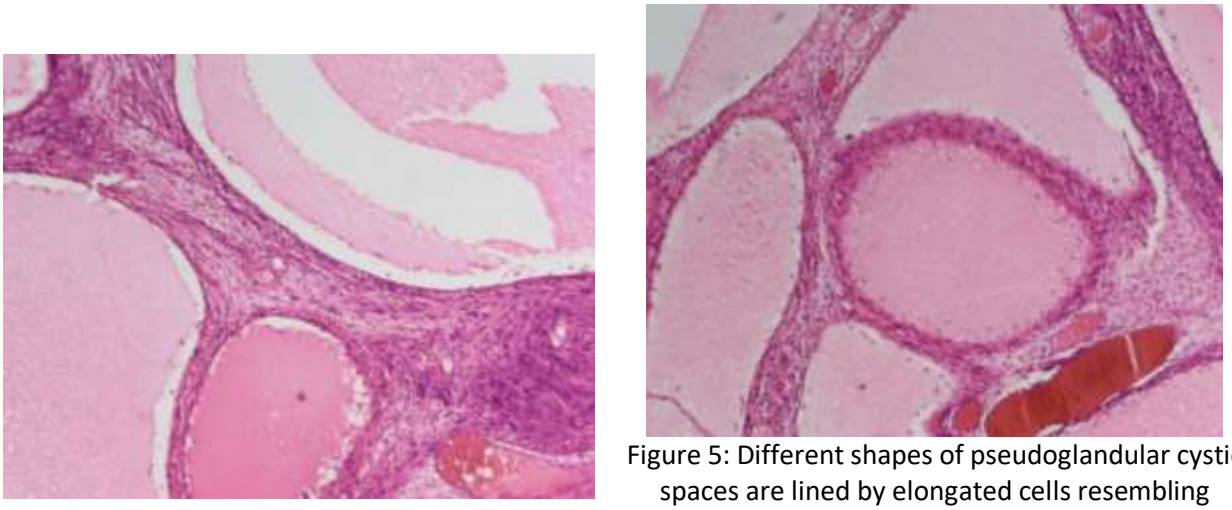

Figure 5: Different shapes of pseudoglandular cystic spaces are lined by elongated cells resembling pseudostratified columnar epithelium, cuboidal to flattened cells (H&E x 400) (A-B),

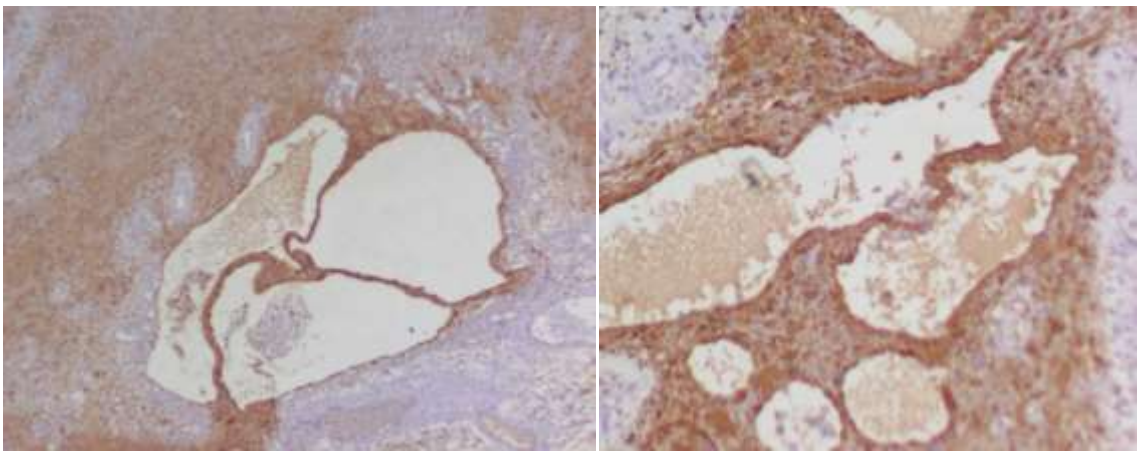

Figure 6: S-100 protein positivity in the cells lining pseudoglandular cystic spaces as well intervening cells (IHC x 100)

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Discussion: Schwannomas are benign tumors, which arise from Schwannoma cells. These are present throughout the body, but more common in head and neck. Schwannoma appear histopathologically as well circumscribed and encapsulated lesion, with biphasic pattern composed of AntoniA&Antoni B areas with occasional palisading (verocay bodies). Swedish neurologist Nils Antoni first described the entity in 1920 and is named after him[7] . In the current study, we describe the clinical and pathological findings of Schwannomas received in our tertiary care orthopedic institution over a period of seven years.

Maximum cases were in the age group of 41 – 50 years which is similar to literature[1,2,6,8,9].

Although there is no known gender predilection [2], in the present study, men were affected more than female. Male: Female ratio 2.6: 1, similar to other Asian studies [1,6, 8, 9].

Majority of the Schwannoma were in the spine compared to other studies as our hospital is a tertiary government orthopedic hospital with spine department. In spine, higher incidence is seen in lumbar region 5/22 similar study by Joe HoJeon et al [9]. 70 – 80 % of the schwannomas are Intradural, extramedullarly [9] in the literature and is similar to our study.

Schwannoma are very rare in nasal sinus. They present as nasal polyp. In the literature nasal Schwannoma have been reported as 4% of head and neck schwannomas till date. We had a case of sinonasal Schwannoma in a 38 year old male. It has distinct features as it was unencapsulated.

Clinical features:

Schwannoma are slow growing tumors. They frequently appear as asymptomatic lumps or incidental findings on imaging.

Sensory complaints such as radicular pain and motor signs are common in the intraspinal growth of spinal Schwannoma [2].

Most of our patients presented with low back ache, which was progressive. Few patients complained about radiating pain & weakness, one patient complained of numbness of lower limbs, radioculopathy.

Histopathology: Schwannoma are benign usually encapsulated nerve sheath tumors composed of well differentiated Schwann cells. Cellular type of schwannomas was the commonent type 8/22(36.36%) followed by classical, pseudo glandular pattern,ancient, plexiform type. (9.09 %)

Literature search showed findings by Sunita B patil,Chatura KR [6] where classical type was 78.37%, Ancient type was 8.10 %, cellular (5.40) and least common type was plexiform and study by Srikanthshastry[3] in which classical type was majority (82.14%), followed by Ancient (7.14%), cellular (7.14%) and plexiform(3.57%).

Cellular Schwannoma - this type of schwannomas is composed exclusively of Antoni A type and lack Verocay body. Small areas of microscopic necrosis, cellular whorls, perivascular and capsular lymphoid aggregates might be seen [2, 3]. Deeper location of the tumor makes it difficult to do the complete excision. Hence, recurrence rate is high about 40%.

Ancient type of Schwannoma show degenerative changes. It includes myxoid, fibrotic areas, atypical cells, hemosiderin macrophages with predominance of either Antoni A, or Antoni B areas [2]. It is thought that the lesion's long history may have contributed to evolution into an ancient variant [1,3,].

Plexiform Schwannoma are the distinctive subtype. These typically develop in superficial site (skin on subcutaneous tissue) and is characterized by an intraneural nodular growth pattern[9]. There are two types – Biphasic plexiform, cellular plexiform schwannoma. Cellularplexiforn pattern are composed of solid nodules (Antoni A) separated by thin fibrous

bands. These tumour lack well formed capsule & thick walled blood vessels. A marginal connection with NF-2 exists [2]. We had two cases of plexiform Schwannoma. One in tiny finger and another is cervical region that is superficial location as per literature [2].

Schwannoma with pseudo glandular element. It is a same variant in which cystic spaces or gland like structures are formed lined by neoplastic Schwann cells. Flat, cuboidal or columnar cells line these spaces which may contains secretion like eosinophilic substance. These cystic spaces are scattered in Antoni A or Antoni B areas. The lining of these cystic spaces demonstrate positivity for s -100, negative for cytokeratin or EMA.

In a case of sino nasal schwannoma, capsule was not seen. This is similar to the literature[8] .The cause of the absence of a capsule at such mucosal sites are unknown but may be connected to the origin of autonomic neurons that lack epineurium or biological aspects of submucosal setting[8].

Conclusion

Schwannomas are rare benign neoplasm that typically appears as a single, solitary asymptomatic lump anywhere on the body. Schwannoma should be considered in the differential diagnosis of any slow growing asymptomatic lesion. It has a wide range of morphological variation and offers a challenge in the diagnosis. Understanding the morphological variants of schwannomas aids the pathologist in making a precise diagnosis.

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