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RESEARCH ARTICLE

HISTOPATHOLOGICAL STUDY OF PERIPHERAL NERVE SHEATH TUMORS.

Ghatge R. M, More S. S, Gupta. S, Inamdar A. A, Verma A and Sujata. N.
Post graduate student, Department of Pathology, Dr. D.Y. Patil Medical College, Kolhapur.

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Abstract

Accurate pathological designation of peripheral nerve tumors is important as there is dramatic difference in clinical outcome between benign and malignant tumors.Objective was to classify peripheral nerve sheath tumors as benign and malignant and to study the distribution of peripheral nerve sheath tumors among different age groups, sex and different anatomical locations. The study includes patients clinically diagnosed and treated for peripheral nerve sheath tumours from Dr. D. Y. Patil Hospital and Research center, Kolhapur. Sample size**: 50** cases. Haematoxylin and Eosin stained sections were examined microscopically and histological interpretation were done. IHC was performed as per requirement. This study was conducted between May 2014 to may 2016. 49 out of 50 cases of peripheral nerve sheath tumors were benign, occurring more commonly than MPNST.PNSTs showed almost equal predilection for both the sexes. PNSTs occurred over a wide age range most common between 2nd and 4th decade of life.

Introduction:-

Peripheral nervous system – can be defined as nervous system outside the brain and spinal cord. It is an extensive system which includes somatic and autonomic nerves, end-organ receptors and supporting structures.[1]

Schwannoma is a benign, non-recurring tumor arising in cranial nerves, bone, and gastrointestinal tract but mainly in subcutaneous tissue and muscles, with a slight predilection for distal extremities or head and neck region. Histological, is an encapsulated neoplasm having two components, known as Antoni A and B tissue, in variable proportion.[2]

Neurofibroma unlike schwannoma, the neurofibroma is an unencapsulated nerve sheath lesion that may occur in the following forms: (a) a solitary localized nodule;
(b) a diffuse thickening of the skin and subcutaneous tissues; or (c) a “plexiform” tumor, a wormlike, multinodular growth within major or minor nerves. Although only the last form is characteristic of neurofibromatosis, that autosomal-dominant disease may also produce solitary or diffuse lesions. The neurofibroma may be found virtually anywhere in the skin or subcutaneous tissues, and it is usually seen in young adults.[3]

Malignant peripheral nerve sheath tumors (MPNSTs) comprise a group of tumors that arise from peripheral nerves or that display differentiation along the lines of the various elements of the nerve sheath, including Schwann cells,

Corresponding Author:- Ghatge R. M.
Address:- Post graduate student, Department of Pathology, Dr. D.Y. Patil Medical College, Kolhapur.

perineural fibroblasts, or fibroblasts. Most of these tumors arise on the trunk, extremities, or head and neck regions; they are very rarely located in the abdominal cavity.[118.]

Malignant peripheral nerve sheath tumors (MPNST'S) of the head and neck comprise 2% to 6% of head and neck sarcomas.[4] Approximately half of these tumors arise de novo, and the other half from nerves involved by neurofibromas as part of type 1 Recklinghausen disease.[5]

Our study focuses on most common primary neoplasms of peripheral nerve and emphasizes the histologic attributes of these benign and malignant nerve sheath tumors, to be familiar with these entities and establish their accurate pathological diagnosis in view of their varying biologic behaviour.

Methodology:-

The present study was conducted in the Department of Pathology at Dr. D. Y. Patil Medical College, Hospital and Research lnstitute, Kolhapur . This is a prospective study conducted for a period of 2 years from May 2014 to April 2016. The patients were selected at random irrespective of age, sex, socioeconomic status and residence. The eligible patients were briefed about the nature of the study and a written informed consent was obtained from the selected patients. Findings were recorded on predesigned proforma. Biopsy was performed, specimens were processed with routine technique and Haematoxylin & Eosin staining was performed. Haematoxylin and Eosin stained sections were examined microscopically and histologic interpretation was done.IHC was performed for malignant case.

Results:-

In present study of the 50 cases, In present study of 50 cases, 49 (98%)cases were benign and 1(2%) case was malignant.

Of the 50 cases, 25(50%) cases were Schwannoma,22 (44%) cases were Neurofibroma,1(2%) case of Perineurioma,1(2%)case of nerve sheath myxoma and 1(2%) case of MPNST. Of the 50 cases 27(54%) cases were found in male and 23(46%)cases were found in females.

PNSTs occurred over a wide age range most common between 2nd and 4th decade of life. Youngest patient being 15 years of age and oldest 70 years of age.

PNSTs showed predilection for extremities especially lower limb followed by head and neck region. Three types of neurofibroma were studied,conventional were 12(55%)cases, plexiform were 3(13%) cases and diffuse were 7(32%)cases.

Three types of schwannoma were studied, conventional were 16(64%)cases,plexiform were 6(24%)cases and ancient were 3(12%)cases.

Table 1:- lesions of peripheral nerve sheath tumors.

LESIONS	NO. OF CASES	PERCENTAGE
SCHWANNOMA	**25**	**50%**
NEUROFIBROMA	**22**	**44%**
PERINEURIOMA	**1**	**2%**
NERVESHEATH MYXOMA	**1**	**2%**
MALIGNANT PERIPHERAL NERVE SHEATH TUMORS	**1**	**2%**
TOTAL	**50**	**100%**

Table 2:- incidence of benign and malignant peripheral nerve sheath tumors

Type	No of cases	Percentage (%)
Benign PNST	49	98%
Malignant PNST	1	2%
Total	50	100%

Table 3:- sex distribution of peripheral nerve sheath tumors

Sex	No of cases	Percentage (%)
Male	27	54 %
Female	23	46%
Total	50	100%

Table 4:- age distribution of peripheral nerve sheath tumors

AGE (years)	NO OF CASES	PERCENTAGE
0-10	0	0
11-20	7	14
21-30	5	10
31-40	13	26
41-50	12	24
51-60	8	16
61-70	5	10
Total	50	100

Photographs:-

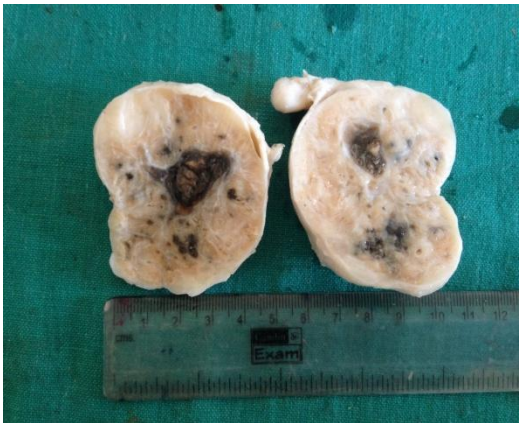

Fig. 1:- Gross specimen of schwannoma

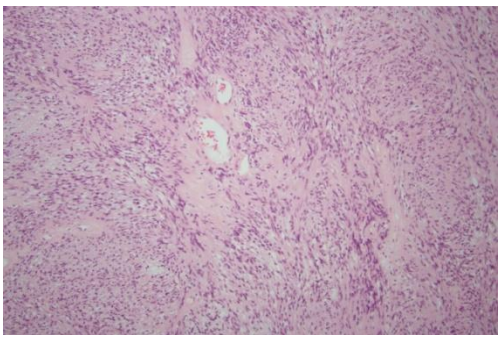

Fig. 2:- Histopathology of schwannoma Photomicrograph showing regimentation of nerve cells along with verocy bodies. (H & E "10X").

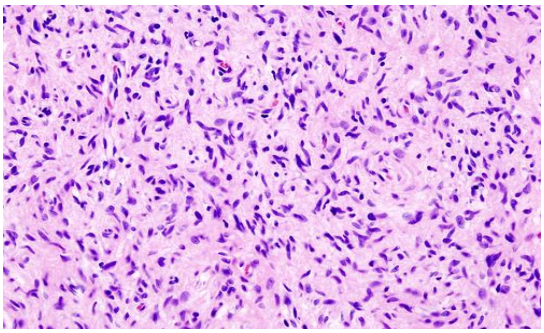

Fig. 3:- Histopathology of Neurofibroma. Photomicrograph showing spindle shaped Schwann cells.(H & E "40X").

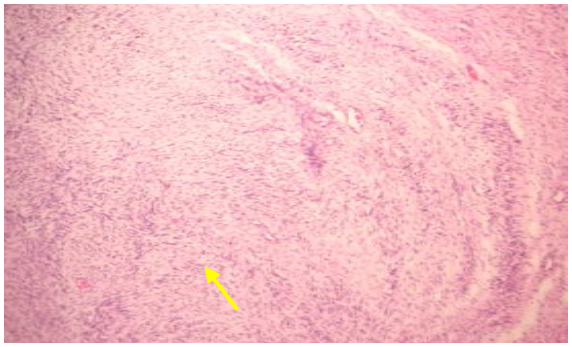

Fig. 4:- MALIGNANT PERIPHERAL NERVE SHEATH TUMOR(MPNST): Photomicrograph showing highly cellular ,mitotically active spindle cells,wavy nuclei with diffuse hyperchromasia.

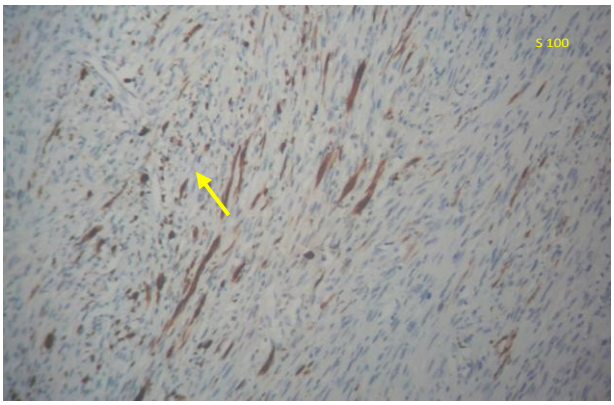

Fig. 5:- MPNST with IHC S100: Photomicrograph showing weak focal S100 staining.

Discussion:-

In our study, out of 50 cases, 49 were bengin & 1 was malignant. According to Casadei et al (1995)[6] and Mohan et al (2003)[7],no of PNST cases were 1256 and25.In our study total no of PNST were 50,out of which 49 were bengin and 1 was malignant

According to Mohan et al (2003)[7],no of PNST were 25 with equal frequency in both sexes.In our study,no of cases were 50 with frequency more in males.

According to Johnson et al(1989)[25] & Mohan et al(2003)[7](1997-1998),neurofibroma were 32 and 10,schwannoma were 27 and 13,MPNST were 9 and 2 cases.In our study of two years neurofibroma were 22,schwannoma were 25 and MPNST was 1 case.

According to Jordan et al (2003)[8]and in our study neurofibroma were 28 and 22,schwannoma were 18 and 25,ancient schwannoma were 1 and 3,MPNST was 1 in both of them.

According to Tsutumi et al(1996)[9] , Wei et al(2005)[10]and Tanna et al(2007)[11] no of plexiform neurofibroma cases were 1 with location of submandibular gland,uterine cervix and in neck region.In our study 3 were present on chest,thigh and forearm.

According to Varebeke et al(1996)[12], Beggs et al (1998)[13]and Zurren et al (2003)[14] no.of diffuse neurofibroma cases were 1,2 and 1 with locations of neck,ankle and lower back.In our study 7 cases were diffuse with location of extremities,head and neck and trunk.

According to Woodruff et al(1983)[17] ,Woodruff et al(2003)[18] ,Agaram et al (2005)[16] ,Agarwal et al(2007)[15]and Berg et al (2008) [19] no of plexiform schwannoma cases were 1,6,24 ,1 and 22.In our study it was 6 with frequency equal in both sexes and common locations were extremities head and neck regions.

According to Wantanabe et al(2001)[20] ,Takeuchi et al (2001)[21] ,Mohan et al (2003)[7] ,Zhuo et al (2003)[23] ,Allison et al (2005)[22] and Focchi et al (2007)[24] the no of MPNST cases were 49,23,2,27,5 and 1 respectively.In our case only 1 case was found.

Conclusion:-

Key issue for the pathologist includes distinguishing schwannomas from neurofibromas, and MPNSTs from cellular schwannoma or neurofibromas. The association of each of these tumors with genetic disorders provides a unique window into discovering basic mechanisms of cell regulation and tumorigenesis that may ultimately shed light on the biology of a much wider array of human disease.

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