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Editorial

The Tinted Bambino-Melanotic Neuroectodermal Tumour Epididymis

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Melanotic neuroectodermal tumour of epididymis configures as an exceptionally discerned, benign neoplasm of neuroectodermal genesis. Initially scripted by Krompecher in 1918 as congenital melanocarcinoma of alveolar process of maxilla, tumefaction is comprised of biphasic population of cells denominated as neuroblastic cells and pigmented epithelial cells [1].

Tumefaction was previously denominated as melanotic adamantinoma, melanotic hamartoma, melanotic progonoma, pigmented congenital epulis, pigmented neuroectodermal tumour, retinal anlage tumour or retinoblastic teratoma.

The infrequently discerned neoplasm commonly arises within young infants with mean age of tumour occurrence at 7 months although tumefaction may emerge between 3 months to 8 years. Besides, adult subjects may be implicated [2,3].

Neoplasm is commonly observed within maxilla of infants wherein a female predominance is encountered with female to male proportion of 2:1. Additionally, tumefaction may be encountered within long bones uterus, brain and diverse cutaneous surfaces. Epididymis configures as an uncommon site of neoplastic occurrence [2,3].

Of obscure aetiology, neoplasm is posited to be engendered from the neural crest, possibly on account of developmental defect within evolution of mesonephric ducts, a site associated with migration of neural crest cells. The principally dys-embryogenetic neoplasm appears to recapitulate development of embryonic retina.

Neoplasm may or may not concur with diverse neural crest tumours as peripheral neuroectodermal tumour (PNET) or neuroblastoma [2,3].

Clinically, neoplasm represents as a firm, rapidly progressive tumefaction. Occasionally, tumefaction may be associated with hydrocele. Generally, neoplasm confined to the epididymis may be observed at birth or detected within first year of life.

Tumefaction may invade the lymphatic spaces and may configure a hydrocele [2,3].

Cytological smears depict a dual cellular population comprised of primitive neuroblast-like cells commingled with clusters of enlarged epithelial cells impregnated with melanin pigment. The cellular population depicts lack of cohesion and appears intermingled within a fibrillary substance [3,4].

Grossly, neoplasm appears as a well circumscribed, firm, spherical to ovoid mass confined to the epididymis. Generally, tumour magnitude appears < 4 centimetres. Cut surface appears grey/white and expounds focal melanin pigmentation. Tumour hue is contingent to quantifiable melanin pigment [3,4].

Upon microscopy, tumour appears to recapitulate the retina as developed at 5 weeks of gestation. Tumefaction is constituted of dual population of cells appearing as neuroblastic cells delineating an elevated nucleo-cytoplasmic ratio admixed with epithelioid cells imbued with melanin pigment [3,4].

Generally, the neuroblastic component displays mitotic figures.

Ultrastructural examination demonstrates enlarged cells impregnated with melanosomes and variably mature premelanosomes. Neuroblastic cells are imbued with neurosecretory granules and neurofilaments [3,4].

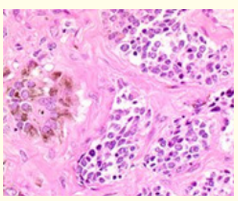

Figure 1: Melanotic neuroectodermal tumour delineating dual population of cells as neuroblastic cells with enhanced nucleocytoplasmic ratio and epithelioid cells imbued with melanin pigment. Surrounding stroma is fibrotic [10].

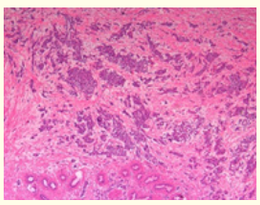

Figure 2: Melanotic neuroectodermal tumour demonstrating dual population of cells as neuroblastic cells with elevated nucleocytoplasmic ratio and epithelioid cells pervaded with melanin pigment. Surrounding stroma is fibrotic [11].

Table 1: Prognostic groups of testicular cancer as per Union for International Cancer Control (UICC) [4,5].

Stage	T	N	M	S
Stage 0	Tis	N0	M0	S0
Stage I	T1-T4	N0	M0	SX
Stage IA	T1	N0	M0	S0
Stage IB	T2-T4	N0	M0	S0
Stage IS	Any T/TX	N0	M0	S1-S3
Stage II	Any T/TX	N1-N3	M0	SX
Stage IIA	Any T/TX	N1	M0	S0
	Any T/TX	N1	M0	S1
Stage IIB	Any T/TX	N2	M0	S0
	Any T/TX	N2	M0	S1
Stage IIC	Any T/TX	N3	M0	S0
	Any T/TX	N3	M0	S1
Stage III	Any T/TX	Any N	M1a	SX
Stage IIIA	Any T/TX	N1-N3	M0	S0
	Any T/TX	Any N	M1a	S1
Stage IIIB	Any T/TX	N1-N3	M0	S2
	Any T/TX	Any N	M1a	S2
Stage IIIC	Any T/TX	N1-N3	M0	S3
	Any T/TX	Any N	M1a	S3
	Any T/TX	Any N	M1b	Any S

Stage IA is comprised of primary neoplasms confined to testis and epididymis. Microscopic evidence of neoplastic vascular or lymphatic invasion appears absent. Clinical examination or imaging demonstrates absence of distant metastasis. Following orchidectomy, serum levels of tumour markers appear within normal limits.

Stage IB is constituted of neoplasms demonstrating localized invasion of primary tumour with an absence of distant metastasis.

Stage IS delineates persistent elevation or enhancing serum levels of tumour markers following orchidectomy, thereby indicating subclinical metastatic disease or a germ cell tumour confined to contralateral testis.

Melanotic neuroectodermal tumour of epididymis appears immune reactive to neuron specific enolase (NSE), synaptophysin, human melanoma black 45 (HMB45) antigen, keratin, vimentin, glial fibrillary acidic protein (GFAP), chromogranin or dopamine β hydroxylase. Besides, special stains as Fontana-Masson may be beneficially adopted to discern melanin pigment.

Tumour cells appear immune non reactive to alpha fetoprotein (AFP), S100 protein or carcinoembryonic antigen (CEA) [5,6].

Melanotic neuroectodermal tumour of the epididymis requires segregation from diverse round cell tumours as rhabdomyosarcoma, Ewing's sarcoma or peripheral neuroectodermal tumour, infiltration from various leukaemia or distant metastasis from neuroblastoma [6,7].

Biochemical examination expounds elevated urinary vanillylmandelic acid (VMA) or homovanillic acid (HVA) levels.

Clinical suspicion of melanotic neuroectodermal tumour of epididymis may be confirmed with cogent histological examination [6,7].

The pre-eminently benign neoplasm may suitably be subjected to surgical procedures as simple orchiectomy [8,9].

Tumefaction is associated with superior prognostic outcomes. Tumour reoccurrence may ensue wherein neoplasms confined to jaw may reappear in ~15% instances. Distant metastasis is exceptional. However, morphological features indicative of tumour reoccurrence or distant metastasis appear to be lacking [8,9].

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10. Image 1 Courtesy: Wikipedia.

11. Image 2 Courtesy: Urology