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Vascular and Neurogenic-Cobb Syndrome

Bajaj A*

Consultant Histopathologist, A.B.Diagnostics, India

***Corresponding author:** Anubha Bajaj, Consultant Histopathologist, A.B.Diagnostics, A-1, Ring Road, Rajouri Garden, New Delhi, 110027, India, Tel: 00911141446785; Email: anubha.bajaj@gmail.com

Mini Review

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Abstract

Cobb syndrome is an exceptional, non-inherited, genetic disorder characteristically constituted by vascular anomalies and neurological deficits. Spinal arteriovenous malformations appear in concurrence with cutaneous vascular lesions within the corresponding dermatome. Dermatome specific port wine stain upon the trunk, arteriovenous malformation, angioma, angiokeratoma, angiolipoma, cavernous haemangioma or lymphatic malformation is discerned in accompaniment with hyperreflexia, limb paresis, muscular cramps, sensory loss, bladder and bowel dysfunction, sudden paraplegia or subarachnoid haemorrhage. Spinal vascular lesions of Cobb syndrome can be adequately determined with magnetic resonance imaging (MRI), computerized tomography (CT) scan, plain radiography or angiography. Cobb syndrome can be appropriately managed with sclerotherapy, endovascular embolization, oral corticosteroids or surgical extermination of vascular lesions.

Keywords: Cobb Syndrome; Magnetic Resonance Imaging; Computerized Tomography

Mini Review

Cobb syndrome is denominated as an extremely exceptional genetic disorder characteristically constituted by vascular anomalies and neurological deficits. Cobb syndrome as a non- inherited disorder delineates a concurrence of spinal arteriovenous malformation in a metamere coincident with a cutaneous lesion. Cobb syndrome can also appear as a component of multiple arteriovenous shunts demonstrating metameric connections [1]. Additionally termed as "spinal arteriovenous metameric syndrome" (SAMS) or "cutaneous meningospinal angiomatosis", Cobb syndrome was initially described by Berenbauch in 1890. However, the syndrome exemplifies a specific clinical setting of spinal haemangiomata appearing in association with cutaneous nevi as described by Cobb in 1915 [1,2].

Disease Characteristics

Cobb syndrome or cutaneous meningospinal angiomatosis enunciates spinal arteriovenous malformations occurring in concurrence with vascular cutaneous lesions within the corresponding dermatome. Cutaneous manifestations of Cobb syndrome appear at birth whereas neurological symptoms emerge around 5 years. Incriminated children lack a family history of Cobb syndrome. Of obscure aetiology, Cobb syndrome probably emerges from somatic mutations within the neural crest or mesoderm with consequent, antecedent, anatomic manifestations within the developing embryo [1,2]. Port wine stain of varying magnitude can be demonstrated on buttock, trunk or thigh. Vascular anomalies may or may not demonstrate a palpable or audible thrill overlying incriminated zones. Vascular aberrations appear blue at birth and fade gradually [1,2]. Cutaneous manifestations of Cobb syndrome vary from macular, port wine stain to diverse categories of papular or nodular vascular lesions such as angioma, angiokeratoma, angiolipoma and lymphangioma circumscriptum. Unlike capillary or cavernous haemangioma, vascular lesions contingent to Cobb syndrome do not resolve or involute spontaneously. Cutaneous vascular malformations are observed incidentally or due to cosmetic concerns or susceptibility to traumatic haemorrhage [1,2]. Commencement of vascular supply to embryological vertebrae and spinal cord emerges from segmental dorsal arteries. Thus, a common metameric origin of arteriovenous blood vessels engendering cutaneomeningospinal angiomas can be contemplated. Vascular cutaneous nevus cogitated

in Cobb syndrome is contingent to several, diverse vascular pathologies. Intra-spinal lesions are usually high-flow, arteriovenous malformations and exceptionally appear as low -flow angiomas. Vascular anomalies with Cobb syndrome can be subdivided into vascular neoplasia such as haemangiomas and vascular malformations [3,4]. Vascular malformations are generally subcategorized as a) slow or low- flow lesions and b) fast or high- flow vascular malformations. Low- flow vascular malformations demonstrate an intermingling of capillary, venous and lymphatic components. The malformations can appears at birth or during early childhood, are gradually progressive and dimensions may increase with crying or adoption of Valsalva's manoeuver. Low-flow vascular malformations are therapeutically amenable to compression garments, sclerotherapy or surgical extermination of the lesion. Cogent therapy can alleviate clinical symptoms of pain, swelling or life threatening vascular malformations as denominated with airway compromise [3,4]. High- flow vascular malformations are comprised of arterial components within arteriovenous anomalies or arteriovenous fistula and can manifest symptoms such as pain, ulceration, ischemic alterations, haemorrhage, congestive heart failure and warm pink patches upon the cutis with accompanying vascular murmur or thrill. High-flow vascular malformations can be adequately treated with surgical eradication of the lesion or embolization [3,4]. Neurological deficits with flaccidity and paresis of upper and lower limbs is enunciated. Deep tendon reflexes or plantar reflexes can be absent. Cobb syndrome is typically discerned following onset of cogent neurological symptoms which appear gradually over a period of weeks or years although a sudden commencement of rapidly progressive muscle paresis can ensue. Neurological representations can vary from monoparesis to sudden onset paraplegia or quadriplegia. Bladder and bowel dysfunction is common although delayed and appears with disease progression. Physical signs such as meningismus, headache, fever, gluteal and limb hypertrophy are infrequent [4,5]. Cord compression due to spinal angioma can induce specific neurological symptoms in addition to activating mechanisms incurring myelopathy such as venous hypertension, compression and cord ischemia arising due to steal syndrome [4].

Clinical Elucidation

Although the disorder appears is late childhood, no age of disease occurrence is exempt. Cobb syndrome represents multiple vascular lesions. Capillary vascular malformations, as designated with port wine stain, demonstrates a dermatome specific distribution upon the trunk. Arteriovenous malformations, angioma, angiokeratoma, angiolipoma, cavernous haemangioma or lymphatic malformations are also delineated. Neurological involvement exemplifies as hyperreflexia with overactive physiological reflexes besides sensory and motor deficits as with muscular paralysis. Morphological or structural anomalies of central nervous system can ensue. Neurological symptoms can appears as intermittent episodes, gradual progressive symptoms or as a sudden onset of pertinent clinical settings. Structural anomalies of the urinary tract can occur. Concordance of a port-wine stain with spinal vascular lesions is a significant feature in accurate discernment of Cobb syndrome and is commonly detected whereas representing symptoms such as pain and motor deficits are non-specific [4,5]. Subjects can depict a sudden onset of limb paresis, muscular cramps in extremities, sensory loss extending up to pelvis besides bladder and bowel dysfunction. Cobb syndrome can demonstrate complications such as a concurrence of kyphosis and scoliosis, bone and joint pain on account of spinal incrimination, vascular thrombosis within arteriovenous malformations, necrotic myelopathy and Foix-Alajouanine syndrome. Exceptionally, congestive heart failure and gangrene can ensue [5]. Symptomatic Cobb syndrome is accompanied with diverse clinical representations and disease course is often unpredictable. Cutaneous lesions can indicate Cobb syndrome, especially when appearing in concurrence with sudden onset of paraplegia or subarachnoid haemorrhage [4-8].

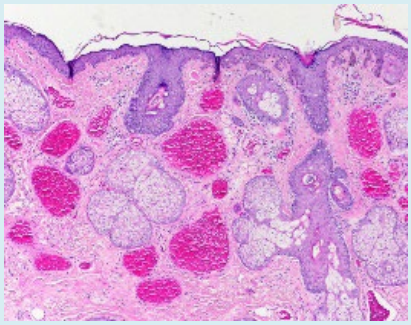

Figure 1: Port wine stain with vascular dilatation, red cell extravasation, cutaneous adnexal structures and a superimposed acanthotic epithelium [9].

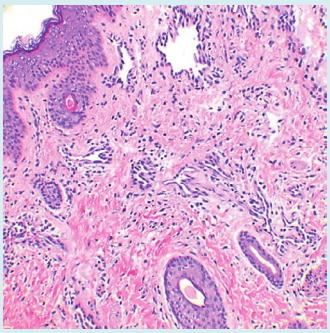

Figure 2: Capillary malformation with vascular distension, encompassing, mildly inflamed fibrous tissue stroma and acanthosis of the superimposed epithelium [10].

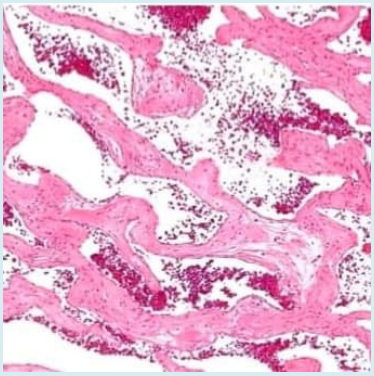

Figure 3: Cavernous haemangioma with enlarged vascular spaces lined with flattened endothelium, red cell aggregation and an intervening, mildly inflamed fibrous tissue stroma [11].

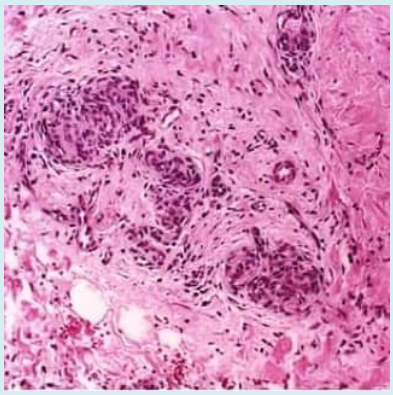

Figure 4: Tufted angioma with vascular spaces lined by plump endothelium with projections, partially solid vascular configurations and an encompassing, inflamed fibrous tissue stroma [12].

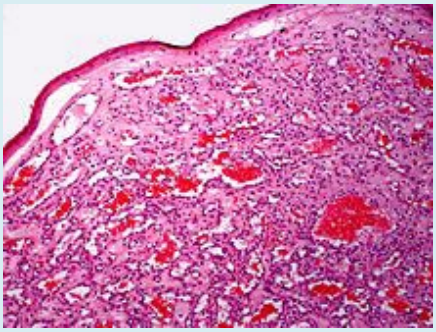

Figure 5: Cherry angioma demonstrating vascular configurations, red cell accumulation, and patent and collapsed vascular articulations and a superimposed squamous epithelium [13].

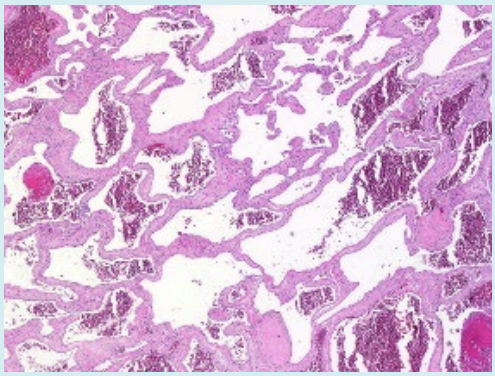

Figure 6: Cavernous haemangioma with enlarged, patent vascular configurations, red cell extravasation and a mildly inflamed, intervening fibrotic stroma [14].

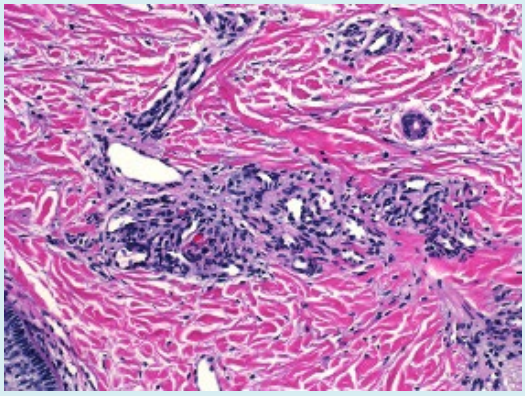

Figure 7: Tufted angioma depicting vascular arrangements lined by plump endothelium with intra-cavitary projections and a circumscribing, inflamed fibrous tissue stroma [15].

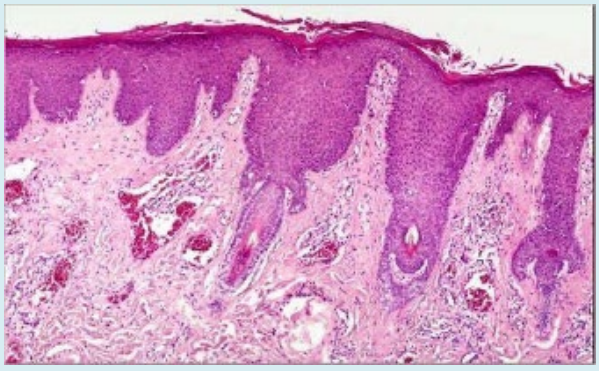

Figure 8: Port wine stain with dilated capillaries disseminated within the upper dermis with a superimposed squamous epithelium showing acanthosis, hyperkeratosis and elongation of rete ridges [16].

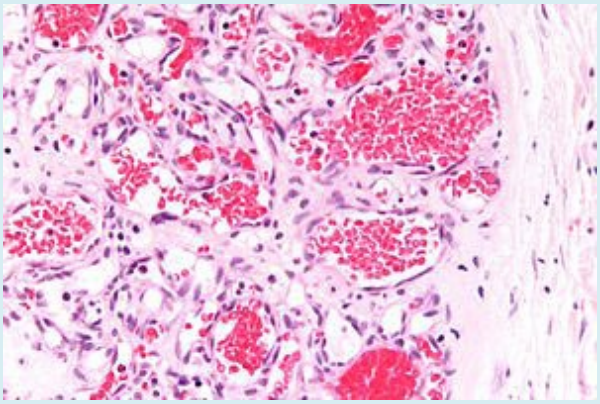

Figure 9: Haemangioma with dilated vascular configurations impacted with red cells and an intermingled, inflamed fibrous tissue stroma [17].

Investigative Assay

Cobb syndrome is discerned due to concurrence of multiple, cutaneous vascular lesions and neurological deficits. Cobb syndrome can be diagnostically confirmed on cogent imaging studies. Lumbar puncture can demonstrate xanthochromia, minimal cell count and a lymphocytic predominance. Proteins are elevated whereas glucose levels of cerebrospinal fluid can be normal [5,6]. Spinal vascular lesions associated with Cobb syndrome can be adequately determined with a magnetic resonance imaging (MRI), although computerized tomography (CT) scan, plain radiography or angiography can also be employed [5,6]. Magnetic resonance imaging (MRI) of dorsal and lumbar spine can reveal an enlarged arteriovenous malformation or haematomyelia within the spinal cord with accompanying haematoma within spinal cord conus. Dilated and tortuous blood vessels can arise from intercostal artery to enter the dorsal spinal canal through cogent neural foramina. Intrathecal vascular enhancement can be discerned upon post- contrast T1 weighted images [5,6]. Computerized tomography (CT) and magnetic resonance imaging (MRI) are advantageous in assessing the extent of lesion. Magnetic resonance imaging (MRI) is superior to computerized tomography (CT) for delineating deformed blood vessels, angiomas and feeding arteries. Ultimate assessment of Cobb syndrome is contingent to angiography. Magnetic resonance imaging (MRI) can demonstrate signal alterations within the intramedullary canal and majority of blood vessels and is contemplated as a superior, safer diagnostic option, in contrast to adoption of an invasive angiography with intravascular contrast. However, selective spinal angiography facilitates exemplification of complex architecture, pathophysiology and adaptable embolization procedures of cogent vasculature [5,6]. Lumbar puncture is contraindicated in the evaluation of suspected spinal lesion especially a spinal vascular configuration [6].

Therapeutic Options

On account of limited discernible instances, definitive management and therapeutic recommendations of Cobb syndrome remain imprecise. Cobb syndrome can be appropriately managed with sclerotherapy, endovascular embolization and surgical extermination of the vascular lesion. Oral steroids can be administered [7,8]. Paraparesis can partially improve and cutaneous nevus can partly regress following administration of oral prednisolone. Endovascular embolization of feeding blood vessels with the employment of n-butyl-2-cyanoacrylate (NBCA) can be adopted, a procedure wherein postoperative neurological deterioration is absent. Continual therapy with oral prednisolone and cogent physiotherapy is advantageous. Supportive therapeutic measures in the form of physical therapy and application of compression garments for capillary, venous or lymphatic anomalies can be adopted [7,8]. Severity of Cobb syndrome is contingent to proportionate incrimination and seriousness of spinal and neurological implication. A preliminary disease discernment and suitable therapeutic intervention is mandated to circumvent the disorder or minimise neurological complications. Optimal management of Cobb syndrome is debatable. Appropriation of a superior therapeutic algorithm can be challenging. Definitive surgical extermination of the lesion is of limited benefit as morbidity due to surgical procedures is of concern. Cogent therapeutic options are palliation without surgical intervention, surgical eradication and a contemporary procedure of endovascular embolization [7,8]. Efficacy of endovascular therapy for spinal arteriovenous malformations in Cobb syndrome requires evaluation. Ambulation with alleviation of extensive paraplegia can be achieved with successful endovascular embolization in enlarged spinal arteriovenous malformations of Cobb syndrome. Spinal arteriovenous malformations are usually extensive and appropriate cure may not be achieved upon multiple, spinal levels. Thus, adoption of combined therapeutic modalities and surgical intervention can induce a cessation of disease progression and minimize neurological sequelae by decimating factors such as mass effect, venous hypertension and enunciation of vascular steal syndrome along the spinal cord [7,8]. Surgical extermination can be beneficially combined with endovascular embolization for treating enlarged spinal arteriovenous malformations. Endovascular embolization can be employed concurrently with pre-embolization steroid therapy. Various treatment modalities can be efficaciously employed with steroids to circumvent disease progression with acquisition of certain degree of neurological function and decline in quantifiable embolization procedures [7,8]. Administration of systemic corticosteroids is efficacious in adjunctive cutaneous vascular malformation such as Kasabach-Merritt syndrome.

Corticosteroids are empirically administered in the exceptionally delineated Cobb syndrome and therapeutic outcomes require further evaluation. Concurrence of endovascular embolization and oral corticosteroids can decimate morbidity and ensure a rapid recovery [7,8].

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9. Image 1: Pinterest.

10. Image 2: Research Gate.

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12. Image 4: Sceilo.br.com.

13. Image 5: Wikipedia.

14. Image 6: Pathology outlines.

15. Image 7: Dermatology advisor.

16. Image 8: Research Gate.

17. Image 9: Libre pathology.